Therapeutic Apheresis Program
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What is sickle cell disease?

Sickle cell disease is an inherited blood disorder; that means it is passed down from a parent's genes. The disease causes the body to make abnormal hemoglobin, which is the protein in red blood cells that carries oxygen to all parts of your body. When you have sickle cell disease, your body tissues and organs do not get enough oxygen due to the abnormal hemoglobin. Healthy red blood cells are round and move easily throughout the body. With sickle cell disease, the red blood cells are shaped like the letter 'C', they are hard, and they are sticky and clump together easily. They cannot move easily through the blood vessels like healthy red blood cells can because they get stuck and block blood flow. This blockage stops the movement of healthy, oxygen-rich blood, and that blockage can cause pain, stroke, vaso-occlusive crisis, avascular necrosis (AVN), anemia, kidney failure, and more.

Red blood cell exchange (RBCX) - Also known as a Transfusion Exchange

RBCX removes blood using an access to a blood vessel (IV needle, port, catheter, AV Fistula or AV Graft), and circulates it through a machine where the blood is separated into each of its components (red cells, white cells, platelets, and plasma). The red cells including the malformed, sickled cells are discarded and are replaced with healthy red blood cells provided by a blood donor. The donor red blood cells are circulated back to the patient with the other blood components through a return access to a blood vessel (IV needle, port, catheter, AV Fistula or AV Graft).

Red blood cell exchange (RBCX) – What to expect?

Before RBCX:

Prior to a procedure, drink a large amount of non-carbonated, non-alcoholic beverages. It is also good to eat a meal prior to the scheduled procedure. 1 to 3 days before your RBCX you will have blood samples taken for a "CBC" and a "Type and Screen." This is important because this is how we order in and match the blood to be used for your transfusion exchange. If you do not have this blood test taken, we will not be able to do your red blood cell exchange.

Please note: It is important to arrive on time for your appointment, as another patient may be booked for treatment after you.

During RBCX:

If peripheral IV catheters are placed, your arms will be propped up on pillows and you may be asked to squeeze your fist to help promote blood flow. You may experience bruising or discomfort where the IVs are placed. If a central venous catheter (CVC) or a port is used instead, you will have full use of your arms. Your vital signs will be monitored by your nurse during the procedure. You may experience some numbness, tingling, light-headedness, nausea, itching or hives. You should notify your nurse of any side effects or symptoms as soon as you notice them.

After RBCX:

After the procedure is completed, you may feel tired for the rest of the day. Make sure you eat and drink plenty of liquids. Monitor your access (IV needle, port, catheter, AV Fistula or AV Graft) for signs and symptoms of infection, which include: fever, chills, pain, redness, swelling, excessive bleeding, pus or drainage around the area. A transfusion reaction can occur during the procedure, or even days or weeks after the procedure. Call a physician for any signs or symptoms of a transfusion reaction, including chest pain, shortness of breath, fever, urine that is dark, Coca-Cola colored.

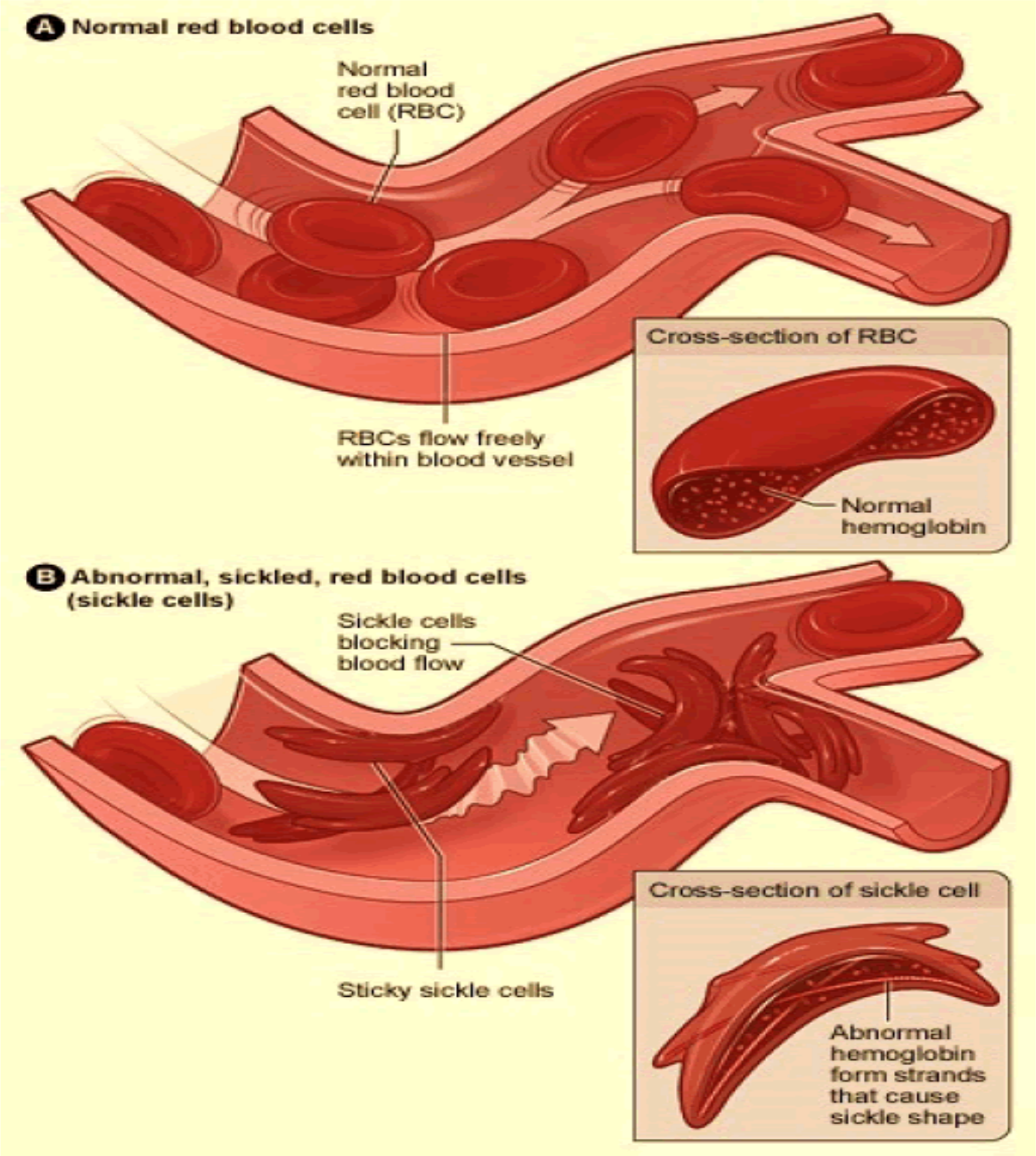
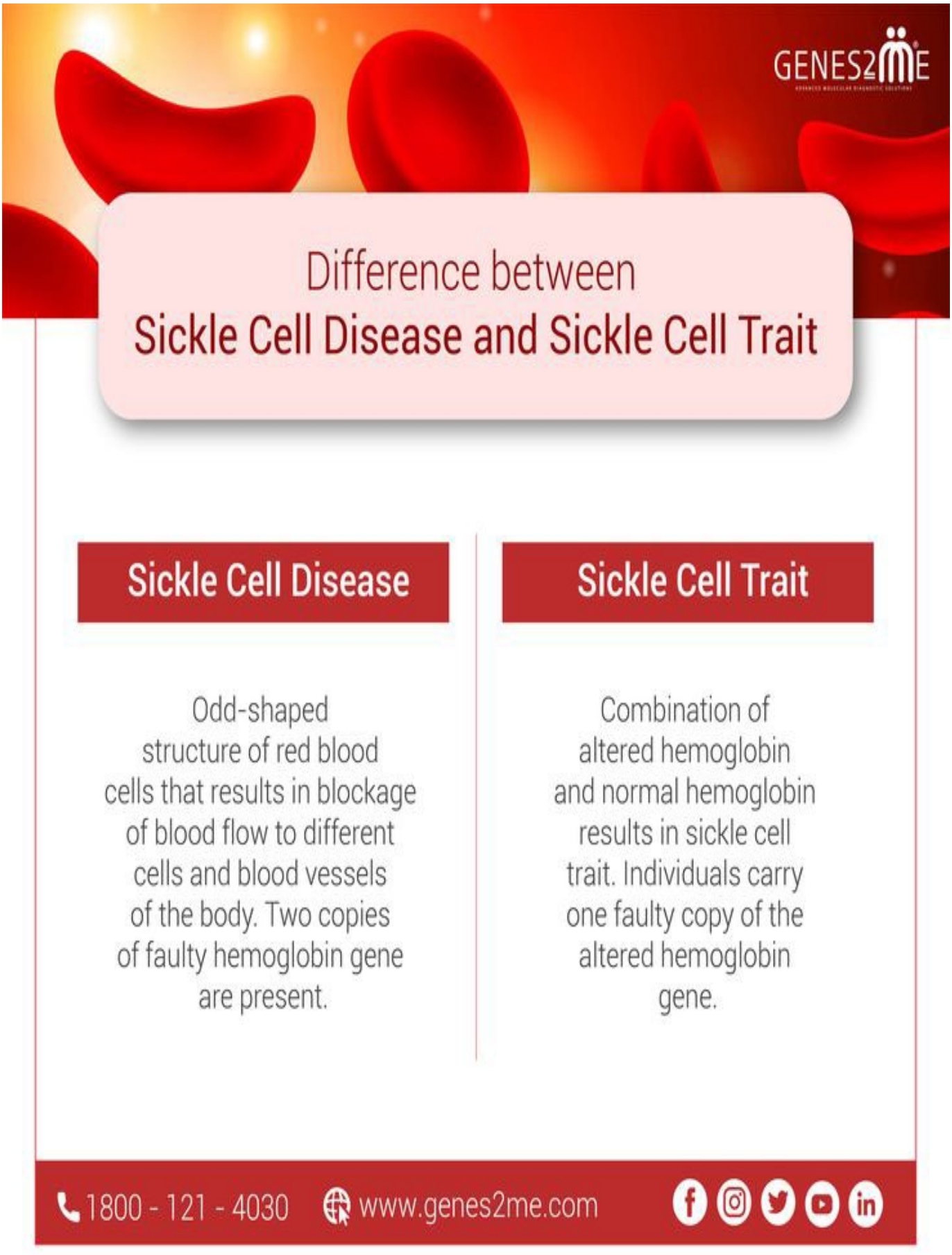


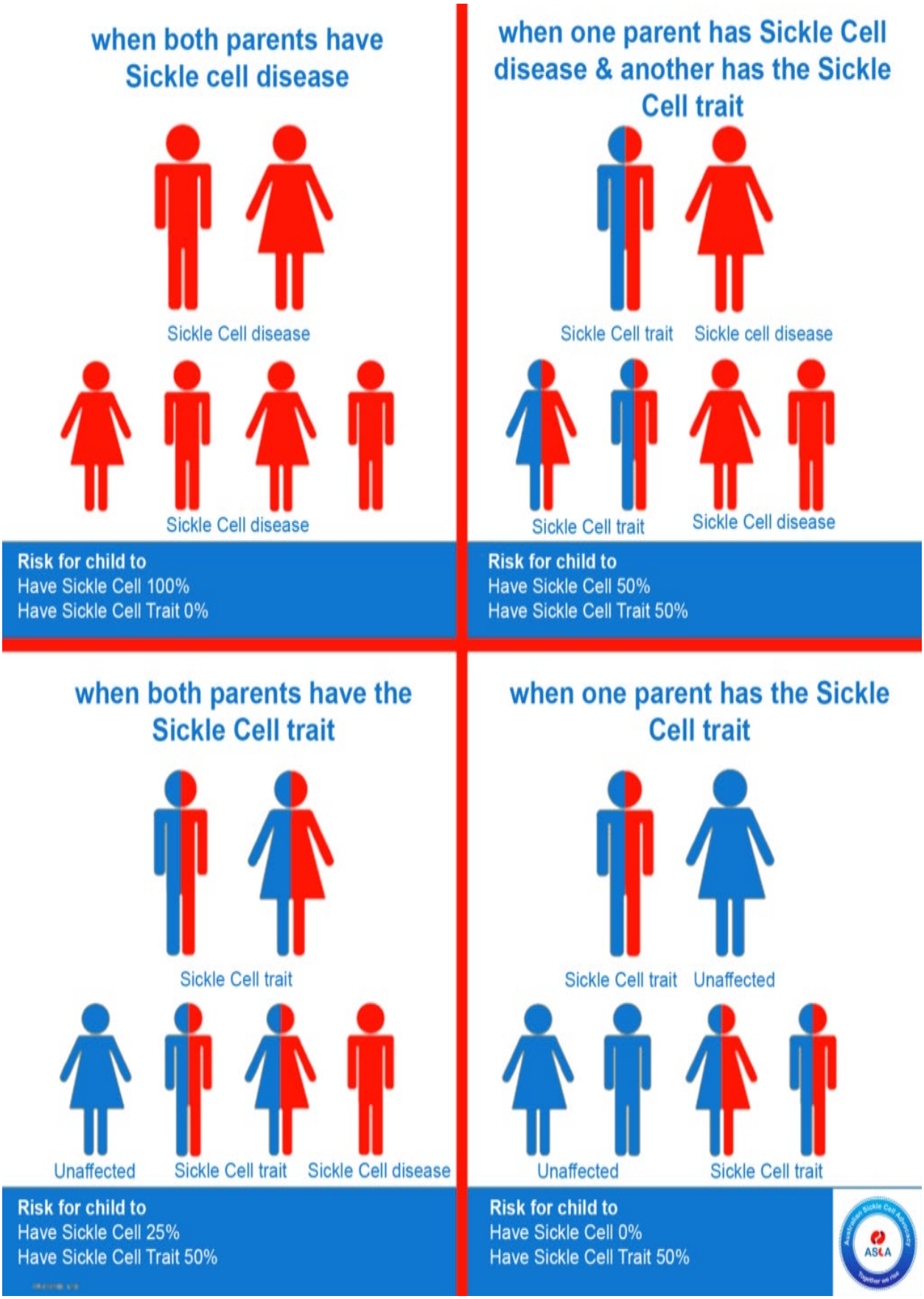
Figure 1: A shows normal red blood cells flowing freely in a blood vessel. The inset image shows a cross-section of a normal red blood cell with normal hemoglobin. B shows abnormal, sickled red blood cells blocking blood flow in a blood vessel. The inset image shows a cross-section of a sickle cell with abnormal (sickle) hemoglobin forming abnormal stiff rods. Source: **http://www.nhlbi.nih.gov/health/health-topics/topics/sca.**

TYPES OF SICKLE CELL DISEASE

Hemoglobin SS Sickle Cell Anemia	Most common type of sickle cell disease (SCD). Inherit one sickle gene from each parent. Regarded as a severe form of SCD.
Hemoglobin SC Sickle C Disease @SickleCell101	Inherit one sickle cell gene from one parent and one 'C' gene from the other parent. Similar symptoms to type SS, less anemia.
Hemoglobin SB + Sickle Beta Thalassemia **Hemoglobin SB 0** Sickle Beta Zero Thalassemia 	The sickle gene produces crescent shaped red blood cells (RBCs) that break down, while thalassemia produces smaller RBCs. There are two types of beta thalassemias. Sickle beta plus thal is mild, while sickle beta zero thal is a severe form of SCD.
Hemoglobin SD, SE, SO Sickle D, E or O Disease	Inherit one sickle cell gene and another abnormal hemoglobin gene (D, E, O, etc.) More rare; the severity of these SCD varies.

SICKLE CELL 101
education + awareness

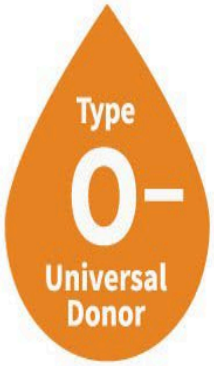

GENES2ME
Difference between
Sickle Cell Disease and Sickle Cell Trait
Sickle Cell Disease
Odd-shaped structure of red blood cells that results in blockage of blood flow to different cells and blood vessels of the body. Two copies of faulty hemoglobin gene are present.
Sickle Cell Trait
Combination of altered hemoglobin and normal hemoglobin results in sickle cell trait. Individuals carry one faulty copy of the altered hemoglobin gene.
1800 - 121 - 4030
www.genes2me.com


when both parents have Sickle cell disease
Sickle Cell disease
Sickle Cell disease
Risk for child to
Have Sickle Cell 100%
Have Sickle Cell Trait 0%
when one parent has Sickle Cell disease & another has the Sickle Cell trait
Sickle Cell trait
Sickle cell disease
Sickle Cell trait
Sickle Cell disease
Risk for child to
Have Sickle Cell 50%
Have Sickle Cell Trait 50%
when both parents have the Sickle Cell trait
Sickle Cell trait
Unaffected
Sickle Cell trait
Sickle Cell disease
Risk for child to
Have Sickle Cell 25%
Have Sickle Cell Trait 50%
when one parent has the Sickle Cell trait
Sickle Cell trait
Unaffected
Unaffected
Sickle Cell trait
Risk for child to
Have Sickle Cell 0%
Have Sickle Cell Trait 50%

Blood Type Compatibility

A blood type is a classification of blood based on the presence or absence of antigens on the surface of red blood cells. Human blood is divided into one of four main blood types: A, B, AB, and O, and is further divided into Rh+ or Rh−.

If you are type	You can receive type: O+	O−	A+	A−	B+	B−	AB+	AB−
O+	●	●						
O−		●						
A+	●	●	●	●				
A−		●		●				
B+	●	●			●	●		
B−		●				●		
AB+	●	●	●	●	●	●	●	●
AB−		●		●		●		●



Blood Compatibility Chart