

Sickle Cell Disease

Blood Disorder | Umbilical Cord Blood



Sickle cell disease is a form of anaemia resulting from a genetic abnormality in the haemoglobin-producing genes, and is usually inherited. It is a recessive disease, meaning that two copies are needed for the disease to be symptomatic. If contracted, the haemoglobin-containing red blood cells form a sickle or crescent shape, which limits the amount of oxygen these cells are able to carry. The red blood cells are also more prone to breakage and to form blockages in smaller blood vessels. Complications of the disease include infections, crises of pain episodes, eye problems, predisposition to strokes and other ischemic events.

Sickle cell disease is more common in people of African and Mediterranean descent, with a single copy being found in 25% of people and between 1% to 2% of all babies born with this disease. It is also seen in people of South and Central America, Caribbean and Middle Eastern descent. It is common for sufferers to die between 20 and 40 years of age. Currently, 12,500 people have sickle cell disease in the UK, with a lifetime cost of between £92,323 to £185,614 per patient.

Clinical Trials

It is known that transplantation of haematopoietic stem cells can treat sickle cell disease successfully. Clinical trial NCT00029380 demonstrates this, with results to be confirmed after follow up. A trial with expanded cells is being planned.

Cell Line

Work has been done to alter the haematopoietic cells from the individual, which in the lab has been shown to be effective. However, these have not been tested in humans, and the expectation is that this is a long way away from clinical trial.

Future Work

Most research is focusing on the symptomatic treatment of the disease and controlling its expression to reduce the severity of the effects in individuals.

Summary

For individuals who are of a high-risk ethnic background, it is acknowledged that storage of cord blood to assist with future transplantations of either themselves or family members should be considered as routine.

References

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