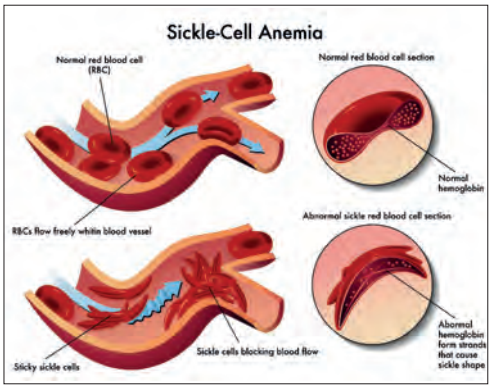
Olea MEDICAL
Improved diagnosis for life™

Did You Know?
Sickle Cell Disease

Take-home message

- Sickle cell disease is a genetic disease, it is not contagious. It is transmissible to a child by its parents.
- Newborns show no sign of illness but from the third month complications may appear.
- Blood transfusions are required for hemoglobin levels below 7 g / dl.



- **Sickle cell disease,** also called **sickle cell anemia,** is a **genetic disease of hemoglobin** (HbS), which deforms red blood cells. This causes severe anemia.
When sickle cell disease is **transmitted by only one parent,** it is called heterozygous or AS: it is **the mildest form.**

When sickle cell anemia is **transmitted by both parents,** it is called homozygous form, or SS: it is **the most serious form.**

Sickle cell anemia is **widespread throughout the world.** It is more frequent in **India, South America (especially in Brazil), Southern Italy, North Africa, Greece** and the **Near East.** In 2013 **441 children** with this disease were born **in France.**

The severity of sickle cell disease varies widely among individuals. Symptoms can occur from the age of three months. Medical care aims at avoiding the complications of the disease.

The available treatments are limited, researchers are trying to find therapeutic alternatives.

Complications

- Chronic anemia.
- Increased spleen volume.
- Child stunting.
- Yellow-coloured skin.
- Painful vaso-occlusive crises.
- Sensitivity to infections.

Possible therapy

- Prevent infections by daily antibiotics and vaccinations.
- Blood flow monitoring by ultrasound or brain MRI to avoid a risk of stroke.
- Blood transfusion that reduces the risk of stroke and restores an acceptable level of red blood cells.
- For very severe forms, the only curative treatment is bone marrow transplantation.

In Olea Sphere®?

Sickle-cell anemia can be investigated by MRI if the risk of cerebral infarction is suspected (clinical signs, high blood flow in Transcranial Doppler in children). The anatomical MR sequences will show chronic lesions (silent infarcts, signs of small vessel lesions) or recent ones.

Olea Vision will **enhance the visualization** and analysis of the circle of Willis (thin-slab MIP reconstruction or 3D MIP) looking for probable stenosis.

Perfusion weighted imaging with contrast agent injection (Perfusion module) or without (ASL module) **may be particularly useful for detecting perfusion anomalies.**

The Relaxometry module (T2* mapping) **can be used to evaluate the iron** overload in the brain parenchyma which may occur as a result of the various transfusion exchanges.

The follow-up longitudinal module can be used to monitor the evolution of cerebral perfusion (during treatment) as well as the evolution of the iron overload.

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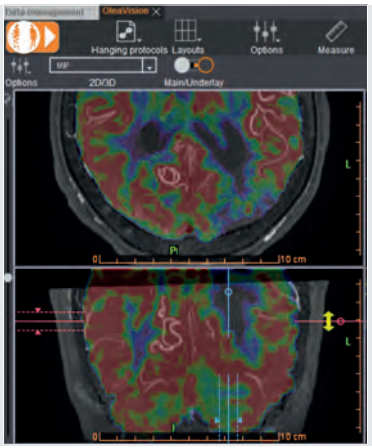
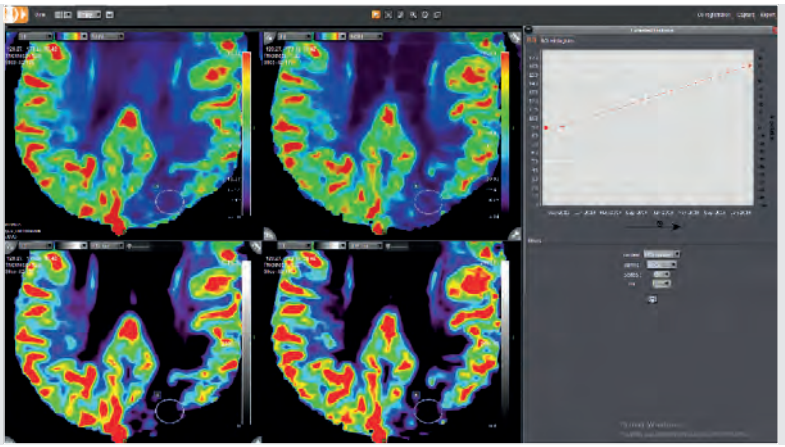
Clinically asymptomatic SCD patients could have WM volume loss with elevated mean platelet volume (MPV), that is a predictive factor of negative outcome. Brain volume measurement and anatomic brain parcellation could be promising tools for asymptomatic SCD patient follow-up.



Picture 1 : Fusion of cerebral blood flow map (rBF) over the circle of Willis reconstructed in 3D MIP for a better evaluation of small vessels



Picture 2 : Follow-up of the blood flow map (rBF) between two MR perfusion examinations of a 36-year sickle cell patient

Sources: ■ http://fr.123rf.com/search.php?word=dr%E9panocytose&imgtype=0&t word=Sickle+cell+disease+%C2%A9&tlang=fr&oriSearch=medicale&srch_lang=fr&sti=m51qbpgg4eyi3m8gww|&mediapopup=16755655 ■ http://www.inserm.fr/thematiques/genetique-genomique-et-bioinformatique/dossiers-d-information/drepanocytose ■ https://www.orpha.net/data/patho/Pub/fr/Drepanocytose-FRfrPub125v01.pdf ■ http://www.vulgaris-medical.com/encyclopedie-medicale/drepanocytose ■ http://acces.ens-lyon.fr/biotic/gpe/dossiers/drepanocytose/html/synthese.htm ■ http://www.afdphe.org/dr%C3%A9panocytose ■ http://www.drepavie.org/drepanocytose_definition.htmf ■ **Ref ■ [1].**Hemoglobin and mean platelet volume predicts diffuse T1-MRI white mattervolume decrease in sickle cell disease patients, Soyoung Choi, NeuroImage: Clinical 15 (2017) ■ **V.A. // N.K. // A.M. // B.T. // S.F**

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