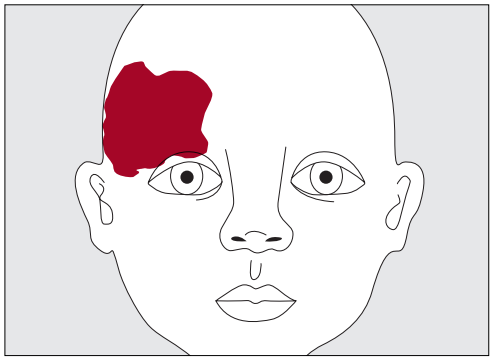
Olea MEDICAL
Improved diagnosis for life™

Did You Know?

Epilepsy in Sturge Weber's Syndrome (SWS)

Take-home message

- The cases of SWS are not identical and the symptoms and severity vary from one individual to another.
- There are cases of SWS where no visible facial angioma is present.
- Patients with SWS develop epilepsy in 75 to 90% of cases.
- Functional imaging (PET scanner, perfusion MRI) is used to evaluate the prognosis.



- **Sturge-Weber syndrome,** also called **neurocutaneous angiomatosis,** is a congenital vascular malformation characterized by **facial capillary malformation, with variable ocular and neurological disorders.**

The cause of this disease is unknown, it is not a hereditary disease and it cannot be cured.

The estimated incidence of SWS is 1 case per 50 000.

Most cases of SWS are generally diagnosed at birth, because of neurological manifestations and the presence of a cutaneous facial angioma (in 87 to 90% of cases). [5]

Epilepsy is a very common symptom in SWS, due to the calcification process.

Patients develop epilepsy in 75 to 90% of cases. **Epileptic seizures in children often appear during the first year of life** and can cause paralysis in some areas of the body and mild to severe mental retardation.

MRI is the exam of choice in SWS screening which may be normal in the neonatal period and must be repeated and **sometimes supplemented by a functional imaging technique in a newborn with a facial angioma.**

Complications

- A facial angioma may extend into areas such as throat, nose, tongue, gums and ear canal.
- Severe epilepsy with uncontrollable seizures.
- Hemiparesis (the slight paralysis of one half of the body, which is opposite to the facial angioma) may also be present.
- Mental retardation.
- Glaucoma (a disease that causes visual loss due to the damage to the optic nerve).

Possible treatment

- Curative treatment of convulsive seizures: medical or, in rare cases, cortical resection; or, in cases of severe epilepsy, a hemispherectomy (surgical removal or deactivation of a cerebral hemisphere).
- The facial angioma can be treated by pulsed dye laser.
- In case of glaucoma, eye surgery may be considered.

In Olea Sphere®?

MR imaging is useful for the diagnosis and evaluation of brain damage in SWS.

In Olea Vision module, MPR (Multi-Planar Reconstruction) **and MIP** (Maximum Intensity Projection) are particularly **adapted to visualize and analyze the data** (Fig 1. et 2.).

T1-weighted gadolinium (Gd) post contrast **MR imaging can demonstrate focal cortical atrophy, contrast-enhancing leptomeningeal angiomatosis (arrow), and abnormal cerebral veins** (Fig. 2).

In Olea Sphere SWI images could be analyzed using **MinIP** projection that **helps to detect the calcifications and veins drainage;** image contrast in **SWI** was increased by using MinIP technique. [1] (Fig. 3)

In **Olea Sphere, Perfusion plugin** (DSC) could show delayed and decreased leptomeningeal perfusion from **TTP** (Time To Peak), **MTT** (Mean Transit Time) and perfusion curve analysis [2] (Fig. 4)

Diffusion and relaxometry modules could also help in SWS diagnosis respectively showing higher ADC in affected area and **increased longitudinal relaxation time T2.** [3][4]

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The SWS diagnosis is done at birth for infants with neurological manifestations and a facial port-wine birthmark in V1 distribution (1/3 upper part of face)

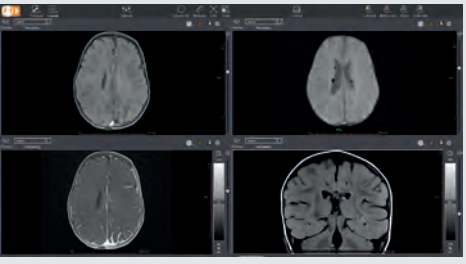

Fig. 1 : Visualization of morphological sequences

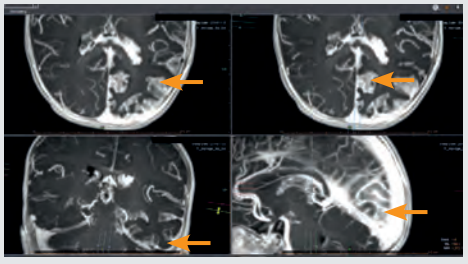

Fig. 2 : . MRI performed on a 9-month baby girl - MIP reformation from 3D T1 post Gadolinium

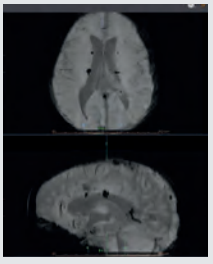

Fig. 3 : MinIP visulization of SWI images.

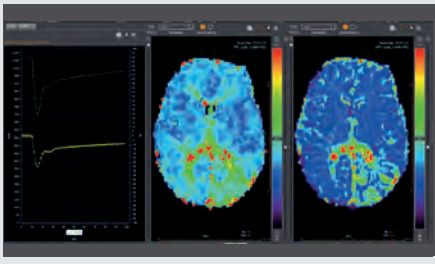

Fig. 4 : Perfusion parametric maps.

Sources: **Ref** ■ **[1]** https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3805373/pdf/nihms507995.pdf ■ [2]: http://www.ajnr.org/content/ajnr/24/9/1912.full.pdf ■ [3]: https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3173907/ ■ [4] http://www.tandfonline.com/doi/pdf/10.1080/02841850510021274 ■ [5] http://pe.sfr-net.org/Data/ModuleConsultationPoster/pdf/2011/1/457dd685-8da8-454d-9173-b1b6b9eac50d.pdf **Other** ■ http://onlinelibrary.wiley.com/doi/10.1111/j.1528-1157.1997.tb00087.x/pdf ■ https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3805257/ ■ https://ghr.nlm.nih.gov/condition/sturge-weber-syndrome ■ https://www.lyceedadultes.fr/sitepedagogique/documents/SVT/SVT_TermS/sophie_codani/T3B0_Rappels.pdf ■ http://emedicine.medscape.com/article/1177523-overview ■ http://www.orpha.net/consor/cgi-bin/OC_Exp.php?Lng=FR&Expert=3205 ■ http://swscommunitycanada.org/fr/sws.php ■ http://www.labome.fr/fr/Sturge-Weber-Krabbe-syndrome-6-cases.html ■ http://www.em-consulte.com/en/article/111770 ■ V.A. // B.T. // A.M. // S.F. // V.I.