ERN RARE-LIVER
European Reference Network on Hepatological Diseases

Sinusoidal obstruction syndrome (SOS)

What is SOS?

Sinusoidal obstruction syndrome, also previously known as hepatic veno-occlusive disease, is a rare condition in which the endothelial cells of the liver sinusoids prolapse into the sinusoid leading to its obstruction. As a consequence the vessel is narrowed, creating an obstruction to blood flow. SOS is caused by the toxicity of certain substances includingchemotherapy, drugs or plants, ...).

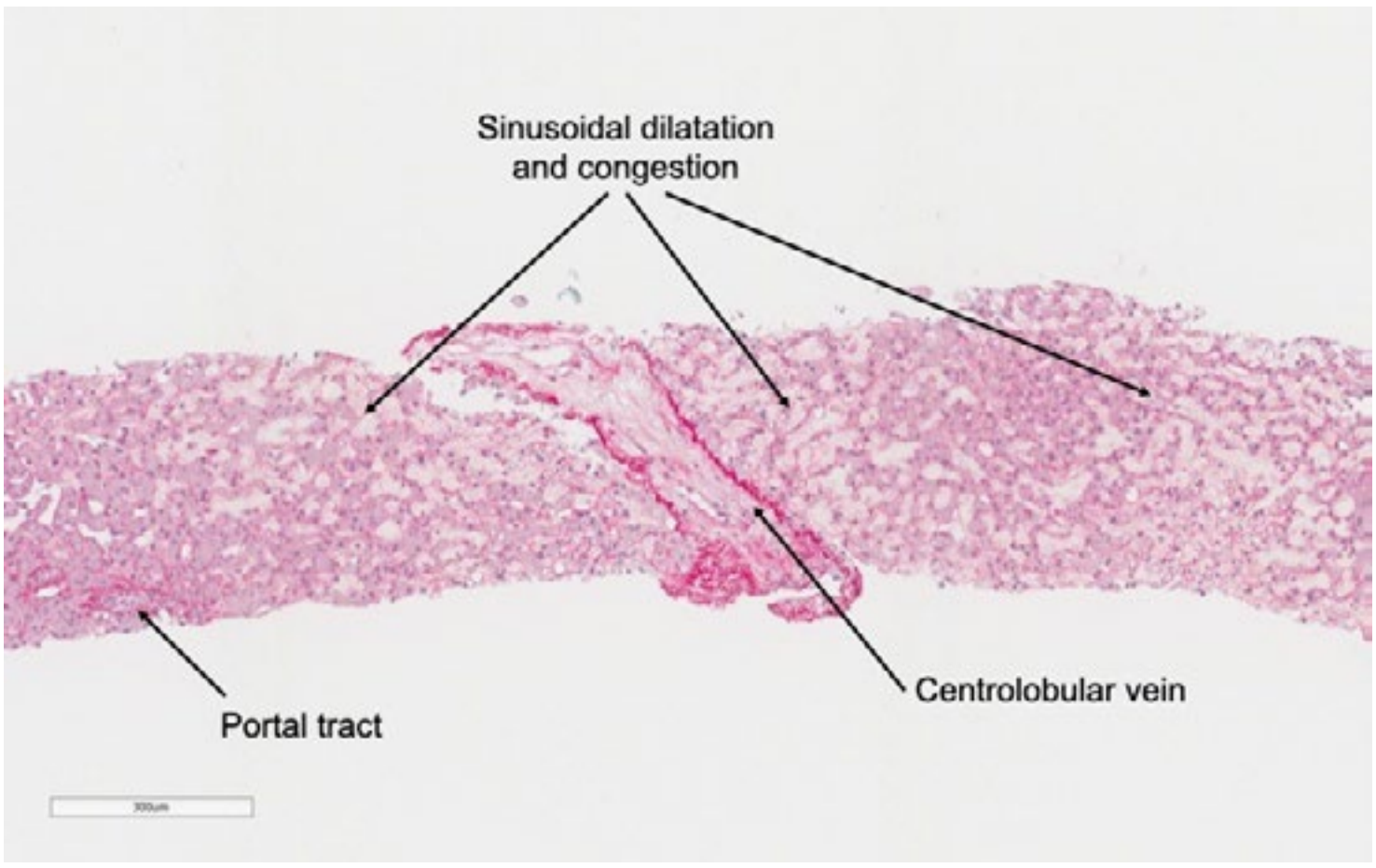
What are the symptoms?

As a result of the sinusoidal obstruction, blood flow out of the liver is impaired.As a result, the liver grows larger, which may result in pain Fluid can accumulate in the abdomen, or in the legs. Both usually are associated with weight gain. Liver function may be impaired. Jaundice, ayellowish pigmentation of the eyes and skin, may occur. In addition, mental status changes called hepatic encephalopathy may occur. Patients may present with a variable number of symptoms, which may vary in severity.

How is SOS diagnosed?

A definitive diagnosis is only possible by measuring blood pressure in the portal venous system and by conducting a liver biopsy. Abdominal ultrasound, liver stiffness measurement and blood tests may be helpful to guide the diagnosis. Excluding other causes of liver disease with blood tests and imaging is also important.

Liver biopsy shows obstructed sinusoids and centrolobular vein.



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Who is affected?

Most cases of SOS occur after hematopoietic stem cell transplantation (HSCT) both in children and adults following chemotherapy and radiation regimens. However, less than 5% of the patients develop SOS after HSCT. Other toxins and drugs are also known to induce SOS, amongst them some immunosuppressive agents, and plant based therapies.

How is SOS treated?

Preventive treatment with ursodeoxycholic acid (UDCA) is recommended for the first period after HSCT. Treatment of SOS is generally symptom-orientated and can include diuretics. In severe cases, Defibrotide, a blood-thinning medication, is indicated. In some cases, when the hematological disease is cured, liver transplantation is a therapeutic option. In selected cases decompression of the portal venous system by TIPS placement could allievate ascites and correct renal failure as a bridge to liver recovery.

What can I do?

Since, in SOS, liver injury is already present, it is essential to prevent further damage to the liver and the kidney by strictly avoiding hepatotoxic and genotoxic agents, such as other specific medications. Alcohol should also be avoided if such a disease has been diagnosed in the past.

Do I need medical checkups?

Yes, it is very important to have regular blood tests, imaging and outpatient clinic checkups by hepatologists, or an experts in SOS management, in addition to the other specialists.

How can I find a specialist?

SOS may need to be managed by a doctor or a hospital network with experience or an interest in SOS. SOS is a rare disease that not every doctor has an interest in or experience with. However, some hospitals are part of the Europe-wide reference network for rare liver diseases, ERN RARE-LIVER. Therefore patients seen at hospitals within the network can benefit from the expertise of specialists who work at other hospitals within the network. For more information about the ERN RARE-LIVER, visit https://rare-liver.eu/. You can find information and patient support in the section “patients” on the ERN RARE-LIVER website (https://rareliver.eu/).

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