

# Susac Syndrome

Patient Review Networking Days, Münster 2023

# Overview

## Disease

1. Process
2. Research



## Patient Care




1. Diagnostics
2. Treatment

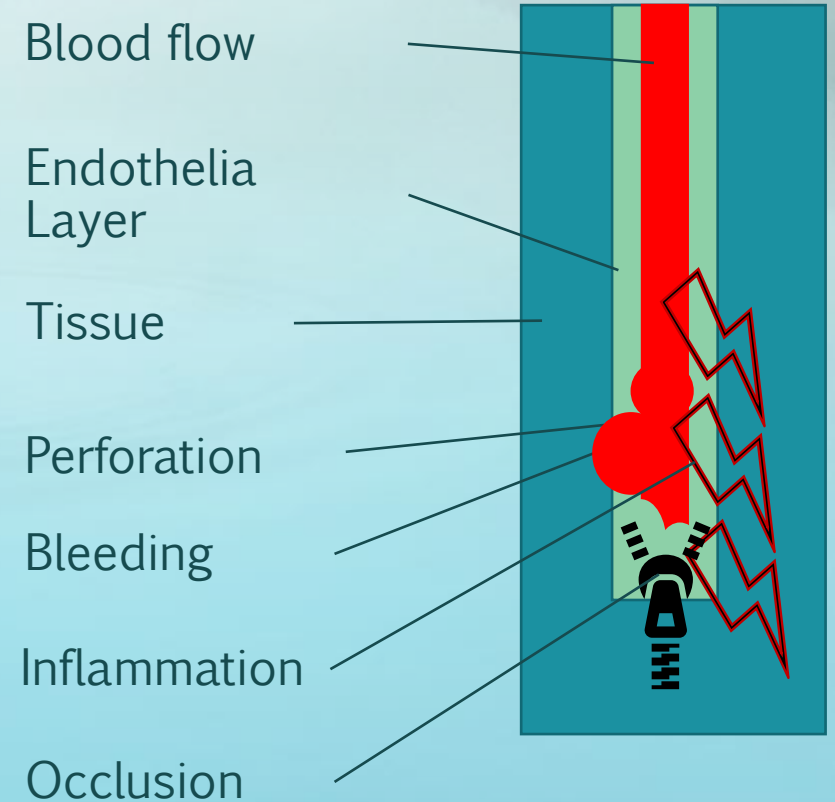


## Conclusion



# Process

- Attack of the Endothelia Cells by CD8 T-Cells
- Inflammation of the blood vessel
- Bleeding into the tissue
- Disturbed provision of the tissue
- Occlusions/ Perforations in the „BEE“ blood vessels
  - Brain (+Spine) 
  - Ears 
  - Eyes 



# Research

- The current research says, that CD8 T-Cells are docking on the endothelial layer in the blood vessels and are expressing Grancyme B and Perforine there. That leads to an inflammation of the blood vessels. This processe causes bleedings:
  1. Occlusions by swelling of the endothelial layer/ bursting of the small vessels
  2. Perforation of the vessel
  3. Bleedings are caused. They are called disturbance oft he blood brain barrier. The blood is irrutting the CNS tissue

This process is happening in brain (and spine), eyes, and ear („BEE“). It was mentioned, that haemagglutinin is only present in the endothelia cells of these organs.



# Diagnosics

**Contrast MRI**  
**Brain and spine**  
Lesions in the Corpus  
Colossum „Snow balls“  
On the roof of the Corpus  
Callosum „String of pearls“  
Lesions can be all over the  
CNS

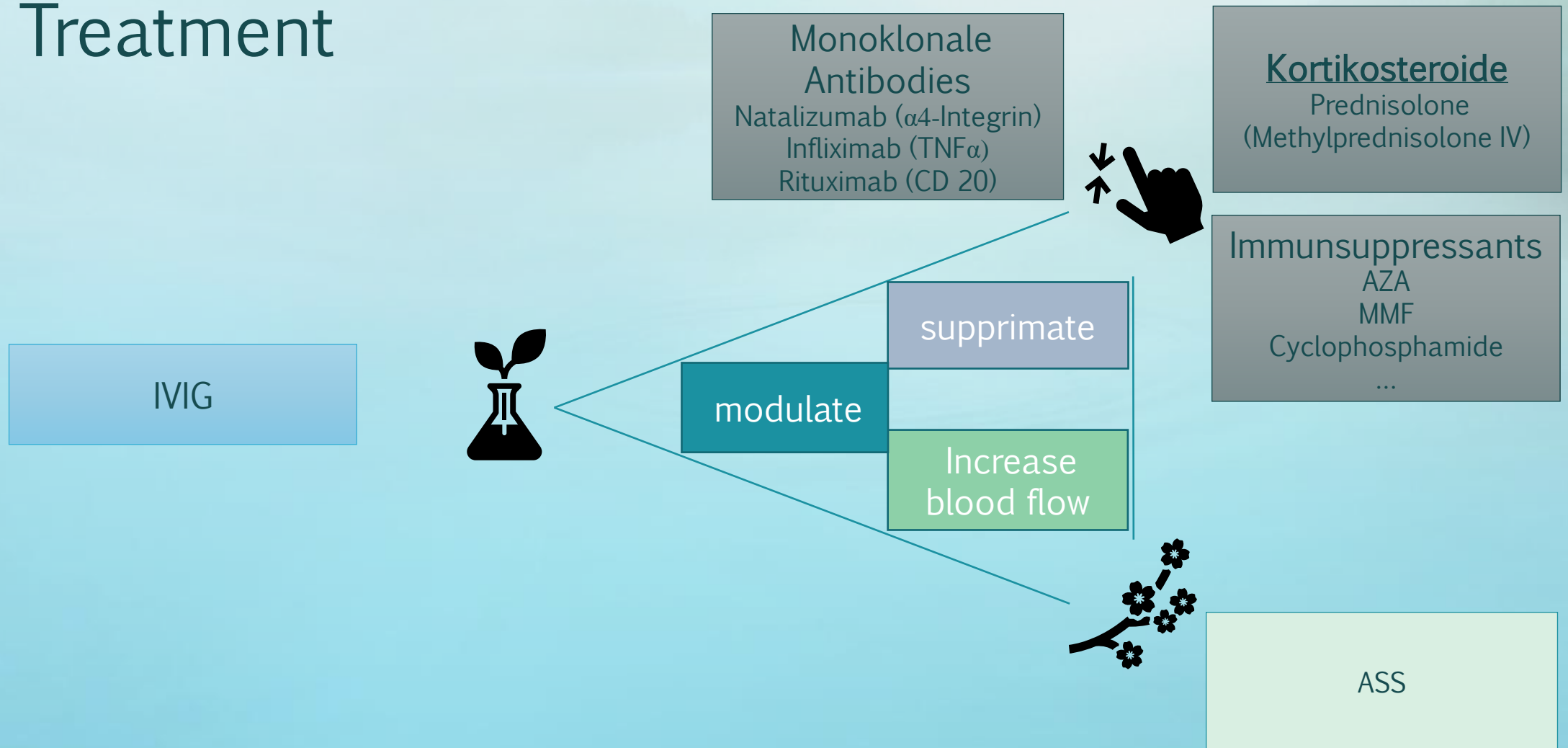


**FA:** Occlusions and  
hyperfluorescating  
areas  
**OCT:** Thinning of  
Susac typical layer  
in the retina

**Audiogramm:**  
middle/low  
frequency hearing  
loss  
**Vestibular tests:**  
VEMP  
Caloric



# Treatment



# Treatment

Methylprednisolone  
Pulses in the  
beginning/ relaps

**Corticosteroids**  
Prednisolone slowly reducing

Immunomodulators  
IVIG

Immunsuppressiva in the long term treatment  
MMF  
Azathioprine  
...

Severe Cases  
Cyclophosphamid

Monoklonale Antibodies in the long term treatment  
Natalizumab  
Rituximab  
Infliximab  
...

Parallel is possible



# Conclusion

- Research on the process of Susac Syndrome is still going on.
- Key is to find the antigen causing Susac Syndrome.
- A worldwide standard in treatment does not exist yet.
- Glucocorticosteroids and IVIG are the basis for further immunosuppressive treatment, especially in the beginning.
- It is unknown if Susac Syndrome is a degenerative disease.
- In most cases, it is assumed that it is only active for a limited period of time.
- A Networking event is helping the exchange, the research and the work on how to handle data. Patients are taken seriously from the present medical Doctors and scientists.

