Epidemiology and Clinical Presentation of Susac's Sydrome in Austrian Population

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Background:

Susac's Syndrom is a rare occlusive microangiopathy of unknown ethiology involving arteries of the retina, cochlea and the brain. The occlusion of arteries in these organs are leading to the typical clinical triad of visual disturbances due to branch retinal artery occlusion, encephalopathy and hearing loss. The syndrome is named after John O. Susac who was the first to described the disease in 1979.

No epidemiological data are available for this disease. Clinical course, treatment regimens, annual incidence and period and point prevalence rates of Susac's syndrome in Austrian population are reported.

Patients & Methods:

All departments of neurology and ophthalmology in Austria where addressed to report adult patients who where on immunosuppressive treatment for the diagnosis of Susac's syndrome between 1 August 2010 and 1 August 2015. The minimum five-year period prevalence rate and the minimum point prevalence rates were calculated in an Austria population of 19 years and older.

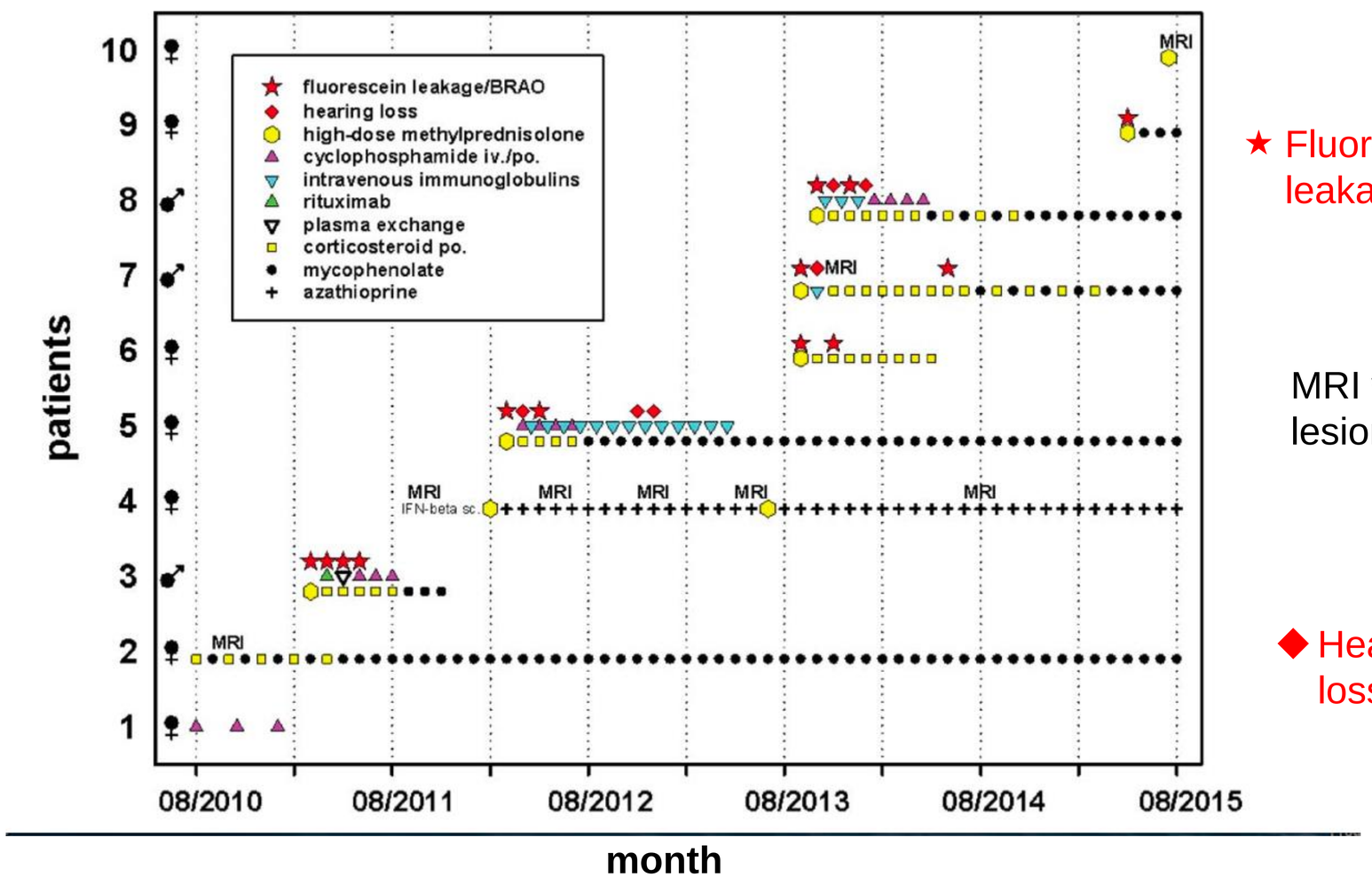
Results:

A total number of 7 departments reported 10 patients (7 women, 3 men) who were on immunosuppressive treatment for the diagnosis of Susac's syndrome between 1 August 2010 and 1 August 2015. Their mean age was 37.1 years (range 22-54) at first presentation and all were Caucasians. On average 6,762,025 people above 19 years of age lived in Austria in the observation period.

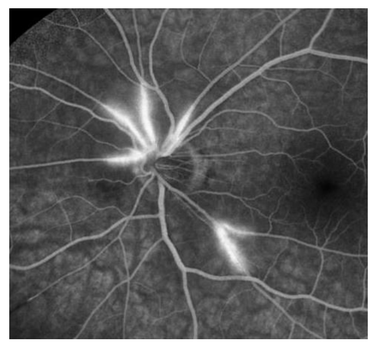
A minimum 5-years prevalence of Susac's syndrome of 0.148/100,00 (95% CI 0.071-0.272) is obtained.

Of all 10 patients 7 (70%) presented with fluorescein leakage and branch retinal artery occlusion (BRAO) initially. Five patients (50%) had hearing loss or tinnitus at first presentation. Eight out of ten (80%) showed central round callosa lesions or internal capsular "string of pearls" on brain MRI, at their first presentation (Fig. 1).

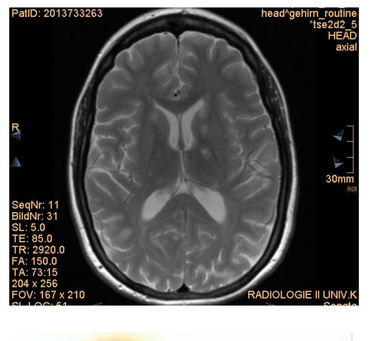
Only 4 out of ten (40%) patients showed the complete clinical triad of Susac's syndrome.



★ Fluorescein leakage



MRI with lesions



◆ Hearing loss

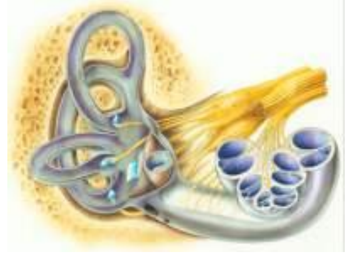


Figure 1.

Clinical course and treatment of all adult patients with Susac's syndrome in Austria between August 2010 and 2015. "MRI" denotes disease activity on brain MRI.

Conclusions:

In this first epidemiological study in patients with Susac's syndrome, we describe the clinical course and provide prevalence and incidence rates in the adult Austrian population. The study serves as a basis for future cooperative research efforts to increase diagnostic awareness and improve treatment of the disease.