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Diagnostic criteria and treatment Algorithm for Susac Syndrome

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Abstract

Susac Syndrome (SS) classically presents with the clinical trial of retinal artery occlusion, sensorineural hearing loss, and encephalopathy and the neuroimaging triad of white matter lesions, deep grey lesions, and leptomeningeal disease. However, cases can present with an incomplete clinical or neuroimaging triad making diagnosis difficult in certain situations. Certain findings on clinical examination or ancillary testing can strongly suggest the diagnosis of SS. These include the Gass plaque in retinal arterioles, arteriolar wall hyper fluorescence remote from retinal vascular injury on fluorescein angiography, and central callosal holes on sagittal magnetic resonance imaging. Recently a new finding, the presence of retinal arterioarterial collaterals, found in the chronic phase of the illness may help in the diagnosis of this disorder. A standard treatment paradigm is also lacking in this illness as a research treatment trial has never been conducted in this disorder. It is important for medical practitioners to recognize clinical and radiographic findings that are pathognomonic for the illness as well as have a basic understanding of the most common treatments in this illness. In this review, I will discuss the myriad of treatments and also how to streamline therapy and adjust treatment as the practitioner strives for remission in their patient.

Biography:

Robert A Egan obtained his medical degree from the Medical College of Wisconsin and completed a Neurology residency and Vascular Neurology fellowship at Oregon Health & Science University in Portland Oregon. He then completed a Neuro-Ophthalmology fellowship at Massachusetts Eye & Ear Infirmary at Harvard Medical School in Boston. He spent the first 9 years of his career at OHSU as associate professor of Ophthalmology, Neurology and Neurosurgery before leaving into private practice. He has remained academic throughout his whole career with a focus on several disorders including Susac syndrome, cerebrospinal fluid hemodynamics, the association of anterior ischemic optic neuropathy related to Viagra, functional vision disorders, and stroke and the eye. He has published over 50 scientific papers, numerous abstracts, and 12 book chapters.

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