

PROSPER MENIERE'S MANY HEADED HYDRA

Current treatments of Meniere's disease can frequently manage the tornado like vertigo, but surgery and medical treatments do mostly little to prevent or stop the inner ear deteriorating. Whilst it is clear Meniere's is multi factorial, progress over many decades rests upon the central idea that Meniere's is a hydroptic distension and ballooning of the endolymphatic space with the central dogma that Reisner's membrane ruptures and causes vertigo. Many authors have speculated on each "cause", with each specialist often steadfast in their beliefs; a viral, vascular, autoimmune, autoinflammatory, obstructive debris, allergic, endocrine, water transport defect or metabolic disorder origins of endolymphatic hydrops have been proposed.

More recently, geneticists have teased out various Meniere's associated genes and have delineated subjects according to symptoms with an emerging concept of Meniere's, AEID, MAV, Cogans and other disorders being a spectrum of syndromes rather than one disease. Despite significant progress, the inner ear is undoubtedly a highly complex organ with unique immunological features and similarly complex metabolic, hydraulic and hormonal mechanisms. It seems that in vulnerable temporal bones, the true nature of Meniere's is akin to a 10 headed hydra, thereby making a one-size-fits-all treatment approach difficult.

Atrophic degeneration of the stria vascularis, alterations to the blood barrier, changes in fenestrated vessels of the endolymphatic sac and ongoing fibrosis and bone remodeling leading to a reduced endolymphatic duct and sac capacity seem consistent characteristics of hydroptic ears. An answer to, "What are these underlying processes that gives rise to the eventual symptoms?" is key to understanding which combination of Dr Meniere's hydra heads rear up in any one individual.

Recent work at the Meniere's Research Fund on an updated Gibson-Arenberg drainage animal model shows attacks of vertigo, at least in the initial stages, are unlikely to be ruptures, but part of a hydro mechanical process that stimulates vestibular end organs when hydroptic distension has reached its limits and endolymph volume alters usual homeostasis in the utricle. With increasing knowledge gained from study of immunological features and immune response of the eye and brain, the inner ear comes into sharper focus with its own unique MALT response and limitations - especially how it deals with exogenous and endogenous antigen, immune complex deposition in basement membranes, damaged endothelial cells and fibrosis formation. Further, fibrotic ears are more sensitive to alterations to the sodium potassium pump where an increase in sodium and reduction in potassium intensifies osmotic pressure and irregular aquaporin activity increases endolymphatic volume. Whether from inconsistent control of endolymph production at the stria vascularis, reduced absorption and processing at the endolymphatic sac or alterations in endolymph composition, it's clear the highly dynamic molecular control of endolymph and inner ear immunological homeostasis is a complex combination and not a stand-alone process.

With more than a century of international and multi- disciplinary research in review, this presentation discusses findings in both animal models and abnormalities in humans with Meniere's. It examines how environment, lifestyle, infection, metabolics and susceptible genetics can increase the risk for immune complex deposition where immunological insults can set up the inner ear for endolymphatic hydrops and the onset of symptoms. In a world of increasingly personalised medicine to treat complex conditions, rather than a single garden path model of Meniere's, looking at the entire landscape perhaps points the way to future treatment options.

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