

THE MECHANISM OF MENIERE'S DISEASE- THE VIRAL HYPOTHESIS

The endolymphatic sac is the guardian of the inner ear. The defence of the hair cells and supporting cells is vital, as these cannot regenerate. A mechanism exists to transport viruses, bacterial remnants, dead cells, or other noxious agents to the endolymphatic sac which is immunologically competent and can destroy the viruses and remove noxious agents off site and away from hair cell harm. This done by activating a mechanism of longitudinal flow of endolymph.

The attacks of vertigo during Meniere's disease are due to faulty longitudinal drainage. The common factor is that the endolymphatic duct is narrow and longitudinal flow is impeded. This can be due to a genetic cause, fibrosis, an otolith narrowing the duct, a spirochete, a tumour of the endolymphatic sac, etc.

During Meniere's disease when longitudinal flow is activated, in the presence of a narrowed endolymphatic duct, the excess endolymph cannot drain to the endolymphatic sac quickly and a situation exists whereby excess endolymph from the cochlea can enter the vestibular portion of the ear and causes vertigo.

One of the initial causes of the excess endolymph (hydrops) may be the result of a viral labyrinthitis. The excess endolymph accumulates initially in the most compliant region of the cochlea, which is the apex of the cochlea resulting in a low frequency sensory hearing loss. As the amount of hydrops increases, eventually the whole cochlea is affected.

The concept is that there is a trigger volume of endolymph that can activate the longitudinal flow mechanism. Small increases of endolymph may be due to factors such as salt loading, infections, stress causing vasopressin release, etc. There is therefore a cluster of attacks of vertigo until the volume of endolymph decreases sufficiently to prevent further attacks of vertigo and a remission occurs.

It is hypothesised that the endolymphatic sac, as an immune organ, has a memory. If the endolymphatic sac realises that another assault on the inner ear is prevalent, it can activate a mechanism which causes an increase in endolymph volume. This activates longitudinal drainage to remove potential noxious agents from the inner ear and to minimise the risk of damage to the inner ear structures.

If the same virus that caused the initial viral labyrinthitis enters the body and circulates in the blood, but cannot be eliminated then the resulting antigens will reach the endolymphatic sac and cause the endolymphatic sac to react to defend the inner ear, even if the virus has not penetrated the inner ear. This will herald the end of a period of remission and result in another cluster of attacks of vertigo.

There may be certain immunological deficiencies which prevent the elimination of the virus in the blood or when it reaches the endolymphatic sac. It is known that persons who have a HLA deficiency are prone to develop Meniere's disease. (The human leukocyte antigen (HLA) system (the major histocompatibility complex [MHC] in humans) is an important part of the immune system and is controlled by genes located on chromosome 6. It encodes cell surface molecules specialized to present antigenic peptides to the T-cell receptor (TCR) on T cells.)

The viral study will collect specimens of blood when a participant with clinically definite Meniere's disease has had an attack of vertigo after a period of remission of at least 3 months. A further specimen will be obtained when the participant has re-entered a period of remission.

The study seeks to analyse blood samples for viruses, which may play a role in the viral pathogenesis of the acute attack in Meniere's – including HSV-1, -2 (Herpes Simplex virus 1/2), CMV (human cytomegalovirus), VZV (varicella zoster virus), EBV (Epstein-Barr virus), AV (Adenovirus).

So far, a total of 13 patients have been enlisted in the study, and a today of 8 blood samples have been collected for analysis by the Charles Perkins Centre Virologists, at the University of Sydney. We are hoping to get an indication of viral type and load in the blood samples, before and after attacks, which may help explain whether viral labyrinthitis is a cause of Meniere's disease.