What You Should Know About Acute Posterior Multifocal Placoid Pigment Epitheliopathy (APMPPE)

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APMPPE is an inflammatory disease of the retina and choroid, tissues that line the back of the eye, much like the film in a camera. The retina is nerve tissue, which converts focused light entering the eye into a signal, which travels to the brain. The choroid is a blood vessel layer, which nourishes the outer half of the retina. Figure 1 illustrates these tissues and their relationship to each other.

**FIGURE 1.**

In the choroid, there are sections of blood vessels called lobules, and each lobule is fed by one arteriole (a small artery). The choroid looks something like a parquet floor because of this lobular structure. In APMPPE, some of the small arteries become inflamed and blocked. The lack of blood supply to the affected lobule causes poor oxygenation and nutrition to the associated overlying outer retina, which causes it to turn a creamy color. The patient notices blurred vision in areas
of the visual field corresponding to these oxygen-starved areas of the outer retina. Figure 2 shows a normal retina. Fig. 3 shows the retina of a patient with APMPPE.

**FIGURE 2. NORMAL**

**FIGURE 3. APMPPE**

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**What Causes APMPPE?**

No one has discovered the cause of APMPPE, although some clues exist. Approximately 25-30% of patients with APMPPE report a flu-like illness shortly before developing the visual problems, leading many to suspect that an unidentified virus is the cause. Blood for antibody testing and many cultures have yet to reveal a convincing candidate virus or bacterium. APMPPE is not just an eye disease, as some patients show inflammation in the spinal fluid, inflammation of blood vessels in the brain, rashes, and joint inflammation. Patients presenting with the eye problems of APMPPE should be specifically questioned to see if these other associated problems are present.
**Who Is Affected?**

Patients with APMPPE tend to be young (mean age 26 years). There is no gender bias, and all races are equally affected. Both eyes are involved in 90% of cases.

**What Can I Expect with this Condition?**

Vision may drop dramatically at first, but usually returns to a good level within several months. Even with return of vision to good levels, there may be blind spots in the field of vision or residual blurring. The cream colored patches in the retina seen early in the disease will turn into variably pigmented scars, which do not change in size over time. Ninety percent of patients have both eyes affected. Rarely, perhaps less than 5% of the time, the condition recurs. Uncommonly, other tissues of the eye may be inflamed and may require anti-inflammatory drops or dilating drops for a short period of time. If brain inflammation, rash, or arthritis accompanies APMPPE, oral steroids may be used for a period of time. The ophthalmologist would work together with an internist to treat these aspects of the disease.

**Diagnostic Work-up**

APMPPE is usually distinctive and can be diagnosed in the office, but sometimes additional testing is needed to distinguish other conditions and look for associated problems. Fluorescein angiography, a test
involving an intravenous injection of dye, allows a photographer to capture images of circulation within the eye. This test is useful for identifying many problems within the eye, not visible to the ophthalmologist during the office examination. The photographs of APMPPE have a characteristic appearance not present in other conditions (fig.4). Some blood work may be drawn to check for infections or evidence of inflammation elsewhere in the body.

FIGURE 4.

If you have questions about this or other retina conditions, please feel free to e-mail Dr. Browning at contact@retinareference.com. If you would prefer to seek further information on your own, about this and other medical conditions, an excellent resource on the internet is the National Library of Medicine. This extensive website includes a search engine for numerous peer-reviewed medical journals and can be accessed at Pubmed.com.