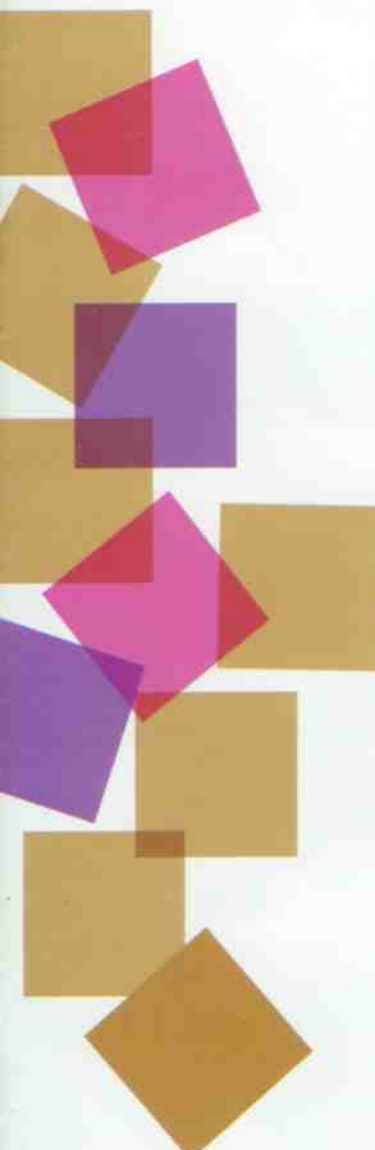


Nataraja Pillai

Oration
December 2005



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Dr. Nataraja Pillai

(1900-2000)

Dr. Subramaniyan Nataraja Pillai was born in March 1st 1900. His medical education was done in Tanjore. He moved to Madras Government Ophthalmic Hospital in 1938, and obtained his ophthalmology diploma in 1939. Col. Wright (authority on Tropical Ophthalmology and who started the Ophthalmology museum) trained him

He continued to serve in Madras till 1943 and shifted to the Government Erskine Hospital (currently Rajaji Hospital), Madurai> He worked with the Government till 1951 and there after private practice. As an eye surgeon he had special passion for community work, with the help of the TVS business family.

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Vitreo Retinal Society - India

Autoimmune retinopathy, CAR and MAR Syndromes

John R. Heckenlively, M.D.

Paul R. Lichter Professor of Ophthalmic Genetics
Director, Visual Physiology Laboratory
Kellogg Eye Center
University of Michigan, Ann Arbor, MI 48105 USA

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Khajuraho, India



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Paul R. Lichter Professor
of Ophthalmic Genetics
Director, Visual Physiology Laboratory
Kellogg Eye Center
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Ann Arbor, MI 48105 USA

Professor Heckenlively was born and raised in Denver, Colorado (the mile high city next to the Rocky Mountains). He did his Bachelor of Arts degree at Oberlin College, Ohio, Medical degree at the University of Colorado, and his Ophthalmology training at the University of Kentucky, Lexington. He did fellowship training in vitreoretinal diseases at the Jules Stein Eye Institute at UCLA, and Medical Genetics at Johns Hopkins. In 1978, he returned to the Jules Stein Eye Institute in Los Angeles, where he was on the full time academic faculty for 25 years. In the Jules Stein Eye Institute he ran the Visual Physiology Laboratory and the Vision Genetics Center. He held the Chair and was the Underwood Family Professor of Ophthalmology at UCLA, and upon joining the Kellogg Eye Center at the University of Michigan in January, 2004, was awarded the Paul R. Lichter Professorship in Ophthalmic Genetics. Professor Heckenlively is Director of the Visual Physiology Laboratory at Kellogg, and heads the Retinal Dystrophy Clinic. He has published five books and 210 articles and chapters.

Autoimmune retinopathy, CAR and MAR Syndromes

Autoimmune retinopathy is one of the most challenging and difficult conditions to diagnose since there are only few definitive tests, currently it is necessary to assemble a variety of information and facts to give credence to the diagnosis. The electroretinogram plays a central role in diagnosing cases of autoimmune retinopathy (AIR). Patients with AIR frequently present with photopsias, night blindness, decreased central vision, and narrowed or scotomatous visual fields; they may be mistaken as having retinitis pigmentosa.^{1,2} Making the situation more complicated, a few retinitis pigmentosa (RP) patients may develop AIR as a complication to their underlying disease (see below). Very often the patient swears that their vision was normal a year before, and now has noticeable changes. On examination they frequently have minimal retinal changes, and many patients are referred to neuro-ophthalmology clinic for evaluation. Most AIR patients develop a diffuse panretinal atrophy. This manifests as a blond fundus, with mild to severe retinal vessel attenuation, and often a fine pigmentation or granularity to the subretinal space. A large majority of AIR patients will not have bone spicule-like dark pigment deposits. The signs and findings in AIR are often subtle and confusing, but an electroretinography *will demonstrate severe retinal dysfunction* in face of often minimal changes in the fundus (Figure 1A and B). Many patients have negative or greatly reduced waveforms in the dark-adapted bright flash ERG. The above findings alone do not give a diagnosis of AIR, but is the first step in establishing a more firm diagnosis.

Autoimmune retinopathy has many variations. Rare patients will have cancer-associated retinopathy (CAR syndrome), and even rarer is melanoma associated retinopathy (MAR syndrome). There has even been a report of

autoimmune retinopathy associated with a teratoma.³ Because different combinations of anti-retinal antibodies have different levels of pathogenicity, and other factors such as blood-retinal barrier integrity, a family history of autoimmune diseases can influence the severity. Circulating anti-retinal antibodies are common in many diseases of the retina, and the challenge is to identify the pathogenic ones from the benign ones.

Most AIR patients present without cancer, but an associated carcinoma needs to be ruled out if the patient has newly diagnosed autoimmune retinopathy. If a patient has a carcinoma or melanoma and then presents with visual dysfunction, the diagnosis is much easier, but still needs to be confirmed with a thorough evaluation, including ERG. Because many of these cases are treatable with immunosuppression that can have significant side effects, it is important to be as certain as possible of the diagnosis.

Autoimmune complications also can occur in patients with retinitis pigmentosa (RP), and the most typical form shows up as severe cystoid edema of the posterior pole or macula. Some patients will have severe striae (wrinkles) of the macular area (not cellophane retinopathy which are rather a mild shimmer effect). These patients typically complain of having noticeable loss of visual field over a short period of time, and if their kinetic visual fields are followed over a year there is noticeable contraction of their isopters every 3-4 months. This subgroup of RP patients have been termed CAR-like syndrome, since they have the similar findings as CAR patients but do not have carcinomas. Most patients fall into the category of simplex RP, but have the additional findings of cystoid edema, retinal striae, diffuse retinal atrophy with minimal to no pigment deposits, and faster progression than typical RP (Figure 2).

CAR Syndrome

Figure 1A. Case 1. 84 year old man found to have Colon carcinoma in Oct. 1994. No vision in OD from advanced glaucoma. Found to have CAR April, 1994. Relatively low doses of Prednisone gave good visual recovery. Larger doses would be used today.

Electroretinogram April, 1994

Photopic
Rod isol.
BrFl Scot

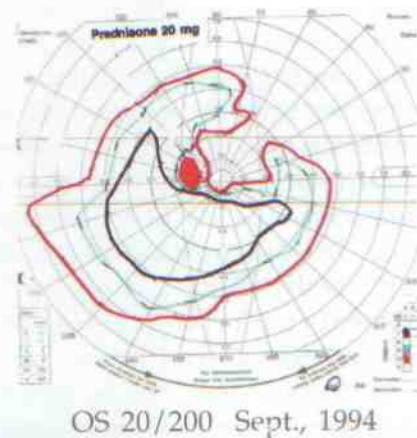
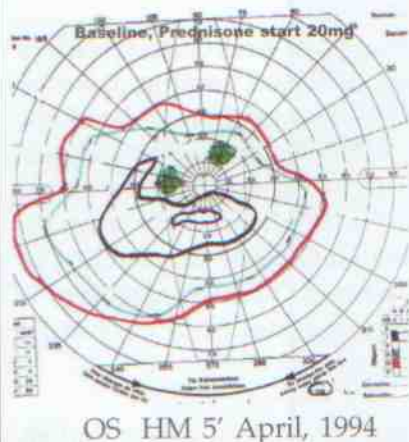
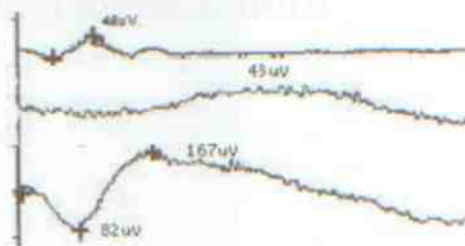
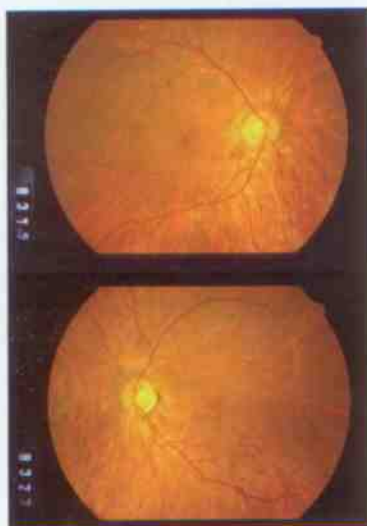
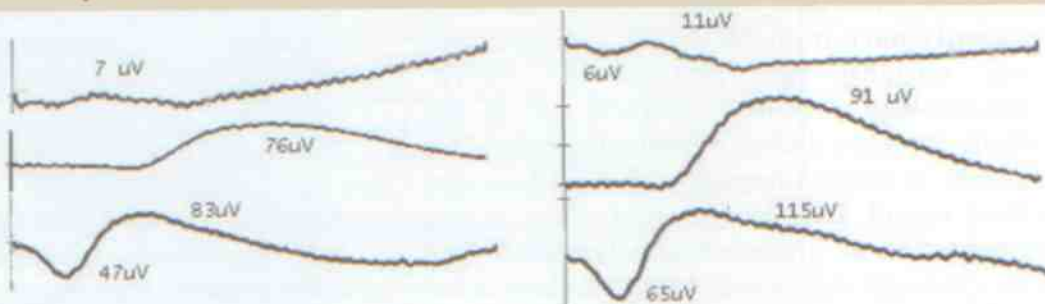


Figure 1B. Case 2. 71 year old lady with ovarian carcinoma found Oct. 2002; vision severely diminished six month later. She was placed on prednisone 60mg, Immuran 100mg, and cyclosporine 100mg. ERG values increased, while Goldmann visual fields remained same on follow up visit. Fundus showed diffuse atrophy without pigment deposits.

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Baseline July, 2003

After Rx October, 2003

