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ACQUIRED PENDULAR NYSTAGMUS IN STARGARDT'S SYNDROME SUPPRESSED BY ALCOHOL

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Stargardts disease is the most common inherited form of juvenile macular degeneration leading to progressive visual loss. We describe a case of female twins aged 47 with Stargardt's Type III (rare autosomal dominant form) who, aged 7, developed blurred vision leading to identical progressive macular dystrophy and deterioration in visual acuities (both 1/60). Pendular nystagmus, presumably secondary to visual deprivation, was observed 4 years ago in both although only one twin (AW) described worsening horizontal oscillopsia, reportedly improving following alcohol consumption. Here, we report measurements of eye movements using 3D video-oculography at baseline and post alcohol ingestion (Blood alcohol concentrations: pre <10 mg/dl, post AW=66 mg/dl and HW=49 mg/dl). Analysis software was used to determine the amplitude and frequency of the pendular nystagmus. In AW peak-to-peak horizontal amplitude decreased from 5° (3.13 Hz) to 0.8°, and vertical amplitude from 2.9° (3.13 Hz) to 0.5°. In HW, peak horizontal amplitude reduced from 4.2° (3.22 Hz) to 1.5° and vertical amplitude from 1.9° (6.25 Hz) to 1.7°. In AW the oscillopsia resolved completely post-alcohol with modified oscillopsia scale scores reduced by 50%. (21/28 to 10/28). The findings demonstrate the potential effects of alcohol in suppressing pendular nystagmus and oscillopsia which may guide pharmacological treatments with alcohol-mimetics.