Hydroxychloroquine Retinopathy

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Abstract
Hydroxychloroquine retinopathy is a cause of irreversible sight loss that can occur in long-term users of hydroxychloroquine. It was previously considered rare, although recent data suggest the overall prevalence is 7.5% after 5 years of therapy, increasing to 20-50% after 20 years. This rise in prevalence is due to the increasing use of hydroxychloroquine for its disease modifying properties in rheumatological and dermatological conditions. Also, the sensitivity of modern imaging technology has enabled detection of retinal abnormalities at a pre-symptomatic stage. Screening for hydroxychloroquine retinopathy is now recommended in the UK by the Royal College of Ophthalmologists.

This study aims to determine the incidence of hydroxychloroquine retinopathy in the United Kingdom. In addition, we aim to ascertain the clinical characteristics of cases as well as the treatment indication. Data will be collected regarding: symptoms, severity of visual loss, risk factors for disease, investigations used to make the diagnosis, any disease progression following drug cessation, management strategies after diagnosis, as well as the route of referral to ophthalmology. This study is timely due to the recent RCOphth recommendations for screening for hydroxychloroquine retinopathy in the United Kingdom.

Case Definition
Hydroxychloroquine retinopathy is defined as bilateral retinal dysfunction (paracentral or pericentral) in a patient taking hydroxychloroquine, in which hydroxychloroquine toxicity is thought to be the principal or sole cause. The diagnosis should be supported by investigations suggestive of hydroxychloroquine retinopathy with at least one of the following tests:

- Automated visual field: central, static visual field (i.e Humphrey visual field)
- Spectral domain optical coherence tomography
- Fundus autofluorescence
- Electrodiagnostic testing (ERG (electroretinography) or Multifocal ERG (mfERG))

Reporting Instructions
Please report any new patient that you have seen in the last month who fulfils the case definition or any patient you have seen in the last month who newly fulfils the case definition. Please include patients referred to you, and patients you have referred to another ophthalmologist. Clinicians reporting cases will receive an initial and a 12-month follow-up questionnaire. Neither the patient nor their family will be contacted, patient management will not be affected, and no special investigations will be required.

Statement of Research Questions
1. What is the incidence of hydroxychloroquine retinopathy in the U.K.?
2. What is the treatment indication in patients with hydroxychloroquine retinopathy?
3. Is hydroxychloroquine retinopathy identified at a pre-symptomatic stage or when the patient is symptomatic?
4. How severe is visual loss in cases of hydroxychloroquine retinopathy (as measured by visual acuity, and visual field mean deviation on 10-2)?
5. What investigations are used to diagnose hydroxychloroquine retinopathy?
6. How is hydroxychloroquine retinopathy managed? Is the drug discontinued, dose reduced, or drug continued?

7. Does hydroxychloroquine retinopathy progress after drug cessation?