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BCR. Birdshot Chorioretinopathy

NIHR BioResource – Rare Diseases study project

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Summary

Birdshot Chorioretinopathy (BCR) is a rare autoimmune condition which is strongly associated with the class I MHC molecule, HLA-A29. The ocular phenotype is of a bilateral posterior uveitis which is seen as a characteristic pattern of chorioretinal lesions (pale yellow flecks or 'birdshot spots'), which may be associated with other signs of mild intraocular inflammation. No clear systemic phenotype has yet been identified. BCR can cause insidious but significant sight loss, and most patients require life-long immunosuppression to try to stabilise the condition.

Due to its distinctive phenotype, BCR potentially provides one of the purest cohorts for experimental studies, in which BCR may be an archetype for the wider group of uveitis syndromes. The Bioresource aims to collect cohorts of patients with BCR for recall to natural history studies, drug trials, and cellular investigations.

Within BCR there is often great variability of course and severity, the cause of which is not known. Collecting cohorts of well phenotyped patients will allow the allelic and trans-acting genetic factors to be investigated, improving our understanding of the biological pathways underlying vision loss and providing opportunities for novel therapies.

Recruitment Criteria

Inclusion is based on the clinical diagnosis of BCR in line with international criteria.[1] In line with these criteria HLA-A29+ is not an essential inclusion criterion, although it is a key criterion for data capture enabling an HLA-A29+ subset of the larger BCR cohort to be identified where necessary.

Inclusion

Patients with a clinical diagnosis of BCR based on the *Levinson et al Research Criteria for the Diagnosis of BCR: Results of an International Consensus Conference* will be included. Required characteristics therefore include: bilateral disease, presence of at least three peripapillary “birdshot lesions” inferior or nasal to the optic disk in at least one eye, with only low grade inflammation elsewhere (no more than 1+ cells in the anterior chamber and no more than 2+ vitreous haze). Retinal vasculitis or cystoid macular oedema are considered supportive but are not essential.

Exclusion

Patients with evidence suggestive of forms of uveitis other than BCR. Specifically the following would lead to exclusion: keratic precipitates, posterior synechiae, or the presence of infectious, neoplastic, or other inflammatory diseases that can cause multifocal choroidal lesions.

References

1. Levinson RD, Brézin A, Rothova A, Accorinti M, Holland GN. Research criteria for the diagnosis of birdshot chorioretinopathy: results of an international consensus conference. Am J Ophthalmol. 2006;141(1):185–7.