

AIV. Autoimmune Vasculitis

NIHR BioResource – Rare Diseases study project

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V1 26/03/2020

Summary

When the body's immune system misfires and damages blood vessels, the condition is called autoimmune vasculitis. Different blood vessels may be affected causing different symptoms and organ damage.



Prof. Ann Morgan, AIV project Lead

Damage to small blood vessels (as occurs in a disease called Anti-Neutrophil Cytoplasm Antibody vasculitis) can result in kidney failure, bleeding from the lungs, and nerve injury. By contrast, when the largest blood vessels are affected in diseases such as in Takayasu arteritis there can be serious damage to the eyes and heart.

All of these conditions have a tendency to relapse following treatment, meaning that patients are subjected to multiple courses of drugs aimed at controlling the disease, and accumulate significant drug-related side effects.

Although we have improved our understanding of the causes of these diseases we have not yet been able to develop sensitive lab tests to help us predict who needs more and who needs less treatment, which is likely to relapse and who will remain in a long-term remission. This information is critical to allow the most appropriate use of treatment for those who need it the most, and avoid potential side-effects in those who do not. Through this project we hope to understand the disease at an individual level, bringing together clinical information about the disease with detailed biological data from genetic and other biochemical analyses. We will make use of patients recruited to the largest European vasculitis database, UKIVAS, and collect through the NIHR BioResource – Rare Diseases study AIV project samples to allow a detailed investigation of a large cohort of patients to take place. This will represent the platform from which new markers of disease activity, predictors of relapse and of cure will emerge.

Recruitment Criteria

Inclusion

Patients with a diagnosis of primary systemic vasculitis including:

(conditions in bold should be prioritised for recruitment)

- **Eosinophilic granulomatosis with polyangiitis**
- **Granulomatosis with Polyangiitis**
- **Microscopic Polyangiitis**
- **Anti-GBM (Goodpasture's) disease**
- Immunoglobulin A vasculitis (formerly Henoch-Schönlein Purpura)
- **Takayasu's Arteritis**
- Polyarteritis Nodosa
- Kawasaki's disease
- **Behçet's syndrome**
- Cryoglobulinaemic vasculitis
- Primary CNS vasculitis
- Cutaneous vasculitis
- Hypocomplementemic urticarial vasculitis
- Other forms of primary systemic vasculitis, including single organ vasculitis
- Unclassified vasculitis

Exclusion

Patients lacking capacity to provide consent.