

INTRODUCTION

Systemic vasculitides are rare, serious disorders which lead to significant morbidity and mortality. Anti-neutrophil cytoplasmic antibody associated vasculitis (AAV) causes a severe systemic small vessel vasculitis often involving lung and kidney.

Microscopic polyangiitis (MPA) and Granulomatosis with polyangiitis (GPA) are 2 of 3 subtypes of AAV with similar underlying pathophysiology. Treatment is with high dose glucocorticoids (GC) and immunosuppressants to achieve remission but relapse is common.

A new product targeted at the underlying pathophysiological mechanisms – neutrophils and the alternative complement system interaction driven through the C5a terminal effector molecule – is being developed. As part of development there is a need to understand the epidemiology of AAV in Europe and how AAV patients are managed in clinical practice since they may be managed by different specialists.

EPIDEMIOLOGY

Pubmed literature searches were performed and existing AAV registries were examined in detail. Epidemiology data were found in centre/regional studies and there is a relative lack of large complete AAV registries across the EU.

Incidence and Prevalence of AAV and its Phenotypes

- **Overall annual incidence rates in Europe:** 10 to 20 cases per million
- **Overall incidence rates from 3 regions in Europe:** Approximately 19 cases per million (similar)
- **Prevalence in Europe:** 46 to 184 cases per million

Incidence of distinct phenotypes of AAV may differ across different geographical regions

- **GPA**
 - More common in Northern Europe
 - **Overall annual incidence:** 11.3 per million
- **MPA**
 - More common in Southern Europe
 - **Overall annual incidence:** 5.9 per million

- AAV can affect people of all ages, but seen more commonly in older age groups:
 - **Peak age group affected with AAV:** 65 to 74 years
- Occurs more often in men (60%) than in women
- Evidence for seasonal variation in incidence for GPA, higher in winter months and a cyclical pattern

Incidence and Prevalence of AAV in Europe



- Relative rarity and nonspecific presentation of AAV are a few diagnostic challenges, which often result in a significant diagnostic delay of more than 6 months in one-third of the patients
- Developing multisystem involvement is often the clue for diagnosis and hence very careful and systematic approach is required for diagnosis
- Diagnosis of AAV depends on the combination of clinical findings and results of imaging, detection of specific ANCA in blood and tissue (often renal) biopsy

PATIENT MANAGEMENT

Essential Principles of Management

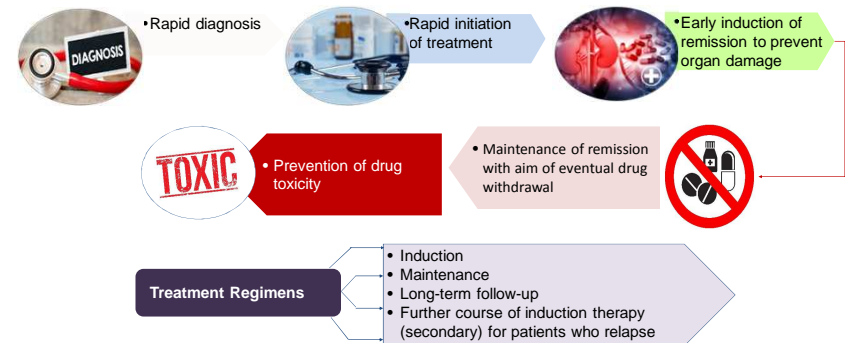


Table 1 – Nephrologists and Rheumatologists are equally involved in diagnosing and managing AAV patients and most AAV treatment is delivered locally (Survey of over 200 physicians, %age response)

	Italy	UK	Germany	France
Nephrologist treats AAV	89	94	94	98
Nephrologist refers patient to another centre	11	3	4	2
Rheumatologist treats AAV	92	97	88	79
Rheumatologist refers patient to another centre	7	2	5	7

CONCLUSIONS

AAV is a rare disease which leads to multisystem involvement and requires careful clinical diagnosis and then long term management to induce and maintain remission. AAV patients are managed by both nephrologists and rheumatologists and most receive care in local hospitals.

Whilst this local service provision carries potential patient benefits, it will be important that all physicians are aware of latest clinical guidance and potential benefits of new AAV treatments to ensure equity of access for AAV patients in the EU.