Real world experience in ANCA-associated vasculitis (AAV) – a complex pathway of patient referral, diagnosis and management

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INTRODUCTION

ANCA-associated vasculitis (AAV) is a severe systemic small vessel vasculitis with frequent renal involvement. Symptoms are variable and delays in presentation to specialist care for diagnosis and treatment are a potential problem. Diagnosis can be challenging and referral pathways to and within secondary/tertiary care complicated. Several different medical specialties, including nephrology are involved in patient management. In addition patient comorbidity is common but poorly reported and may play an important role in acute and chronic clinical outcomes in AAV.

This retrospective study aimed to examine referral, diagnosis and therapy outcomes in AAV patients managed in routine clinical practice in Europe.

METHODS

STUDY DESIGN. Retrospective clinical audit of healthcare records from incident and relapsing AAV patients managed by 399 physicians (240 nephrologists, 120 rheumatologists and 20 internal medicine physicians) who routinely manage incident AAV patients (France, Germany, Italy and UK).

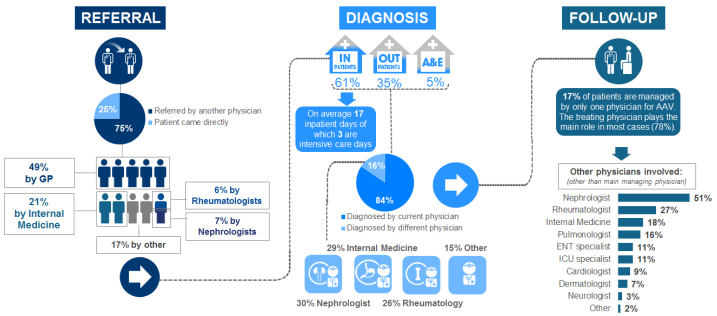
INCLUSION & EXCLUSION CRITERIA. Physicians selected incident or relapsing adult patients with granulomatosis with polyangiitis (GPA) or microscopic polyangiitis (MPA) who had initiated remission induction therapy between November 2014 and February 2017. Patients had at least 6 months of therapy and continuous care by the physician over the time of follow, were over 18 years, had a confirmed diagnosis of AAV for at least 12 months, and had received at least one course of induction therapy to achieve remission.

DATA COLLECTION AND ANALYSIS. Physicians completed up to 3 programmed patient record forms (PRF) - this online data collection tool was designed to gather clinical outcome data over the first 12 months of AAV therapy. Data were collected relating to baseline presentation with AAV then outcomes at 1, 3, 6 and 12 months. Descriptive statistics were used to analyze the data

PARTICIPANTS. 1197 patients were studied in total of which 929 AAV patients were incident patients who commenced remission induction therapy following diagnosis – 54% were classified as GPA and 46% MPA. These incident AAV patients were analysed in detail to describe the referral and diagnostic challenges of AAV.

RESULTS

Figure 1 – Referral and diagnosis. Most incident AAV patients were referred from other physicians, diagnosis was most frequently as an in patient and other physicians were often involved in the diagnosis and subsequent care of the AAV patient.



RESULTS

Figure 2. Symptoms and time to diagnosis Patients presented with a range of clinical features and although renal disease was common, general non-specific symptoms predominated. This may have contributed to the long duration of symptoms prior to diagnosis reported for some AAV patients.

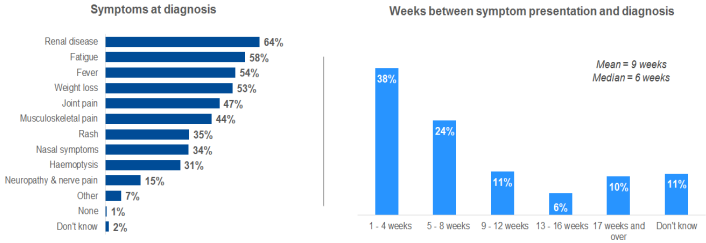


Figure 3. Age distribution The typical gender difference was observed (53.7% male) and AAV incidence was highest in the over 55 age groups.

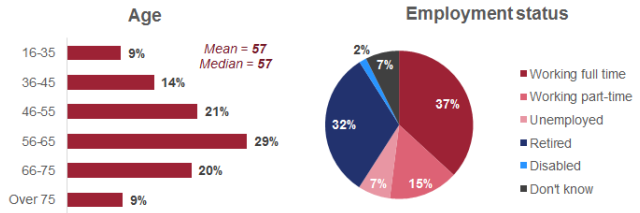
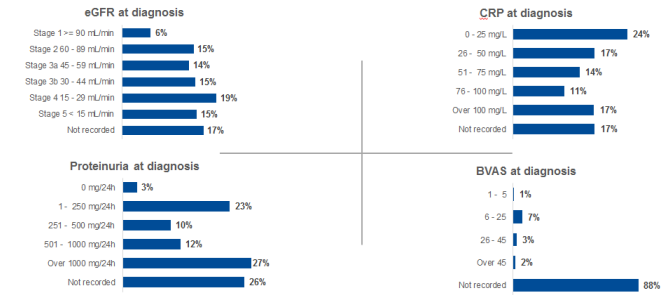


Figure 3 – Diagnostic tests and investigations Serological testing revealed anti-PR3 in 48% of patients and anti-MPO in 47%. Urine abnormalities were common with 62% of patients having haematuria (although 16% of patients had missing values on PRF) and median 24 hour protein excretion was 595mg. Histological support for the diagnosis was performed in 86% of patients with renal histology being the most common (64%) followed by skin (12%) and nose or sinus (11%) biopsy. BVAS was rarely recorded at diagnosis or with patient follow up.



RESULTS

Figure 4 – Disease severity, organ involvement and comorbidity. As BVAS was infrequently reported severity was reported as qualitative scale. Multi-organ involvement was common in incident patients and they also had significant co-morbidity.

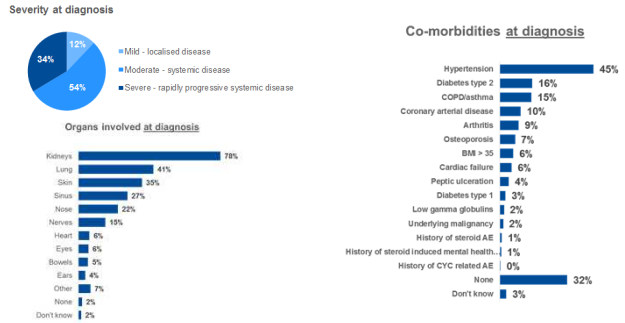
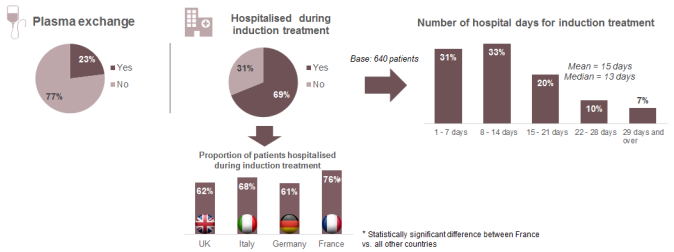


Figure 5 – Interventions and resource utilisation at diagnosis. The majority of incident AAV patients received induction therapy as in-patients resulting in significant resource utilisation. Plasma exchange was used in a significant minority of patients



CONCLUSIONS

This study has used real world clinical practice data from Europe to examine the clinical management of AAV patients.

Incident patients often have a complex pathway to the physician who makes the correct diagnosis and therapy is then delivered in a cross-specialty manner in many cases. Patients are in older age groups and often have renal involvement at diagnosis.

Incident AAV patients typically have multi-organ disease when they present and require induction treatment. Their vasculitis is typically systemic and often severe but formal scoring systems eg BVAS are infrequently used in clinical practice.

Comorbidity is very common in incident AAV patients and the relationships between these comorbidities and the adverse events of the drugs used for remission induction, in particular high dose glucocorticoids, need careful consideration.

Healthcare resource utilisation in incident AAV patients is significant, adding to the unmet medical need in this disease.

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