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 the UK.

METHODS

STUDY DESIGN. Retrospective clinical audit of healthcare records from incident and relapsing AAV patients managed by 100 UK physicians (including 40 Rheumatologists) who routinely manage incident AAV patients

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. 297 patients were studied in total of which 20.5% were relapsing who commenced remission induction therapy following diagnosis – 63% were classified as GPA and 37% MPA. These AAV patients at the time of initial diagnosis were analysed in detail to describe the referral and diagnostic challenges of AAV.

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. Males (49.5%) and females (50.5%) were both affected and AAV incidence was highest in the over 55 age groups.

Figure 4 – Diagnostic tests and investigations. Serological testing revealed anti-PR3 in 52.5% of patients and anti-MPO in 37.4%. Urine abnormalities were common with 61.6% of patients having haematuria (although 15.5% of patients had missing values on PRF) and median 24 hour protein excretion was 365 mg/24 hours. Histological support for the diagnosis was performed in 76% of patients with renal histology being the most common (58%) followed by skin (6.7%) and nose or sinus (5.7%) biopsy. BVAS was rarely recorded at diagnosis.

RESULTS

Figure 5 – Interventions and resource utilisation at diagnosis. The majority of 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 the UK to examine the clinical management of AAV patients. AAV patients often have a complex pathway to the physician who makes the correct diagnosis and symptom duration is often long. Therapy is then often delivered in a cross-specialty manner in many cases. AAV patients are in older age groups and 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 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 AAV patients is significant, adding to the unmet medical need and patient burden in this disease.

DISCLOSURES. This study was supported by Vifor Fresenius Medical Care Renal Pharma a Vifor Pharma Group Company.