of choice for EGPA patients, institution of these therapies in patients with EGPA who are perceived to have severe asthma may delay the diagnosis of EGPA because signs of vasculitis may be masked. Corticosteroids dramatically alter the course of EGPA: up to 50% of those who are untreated die within 3 months of diagnosis, whereas treated patients have a 6-year survival of >70%. Common causes of death include heart failure, cerebral hemorrhage, renal failure, and GI bleeding. Recent data suggest that clinical remission may be obtained in >90% of patients treated; ~25% of those patients may relapse, often due to corticosteroid tapering, with a rising eosinophil count heralding the relapse. Myocardial, GI, and renal involvement most often portend a poor prognosis. In such cases, treatment with higher doses of corticosteroids or the addition of cytotoxic agents such as cyclophosphamide is often warranted. Although survival does not differ between those treated or untreated with cyclophosphamide, cyclophosphamide is associated with a reduced incidence of relapse and an improved clinical response to treatment. Recent studies examining the efficacy of anti-IL-5 therapy with mepolizumab compared with placebo have shown promise, indicating that mepolizumab is a safe and effective corticosteroid-sparing agent that can reduce relapses. Other therapies that have been used successfully in the management of EGPA include azathioprine, methotrexate, rituximab, omalizumab, intravenous gamma globulin, and interferon α. Plasma exchange has not been shown to provide any additional benefit.

## ■ HYPEREOSINOPHILIC SYNDROMES

Hypereosinophilic syndromes (HES) constitute a heterogeneous group of disease entities manifest by persistent eosinophilia >1500 eosinophils/μL in association with end organ damage or dysfunction, in the absence of secondary causes of eosinophilia. In addition to familial, undefined, and overlap syndromes with incomplete criteria, the predominant HES subtypes are the myeloproliferative and lymphocytic variants. The myeloproliferative variants may have acquired genetic abnormalities, including of platelet-derived growth factor receptor α (PDGFRα), attributed to a constitutively activated tyrosine kinase fusion protein (Fip1L1-PDGFRα) due to a