cyclophosphamide, it no longer has a role in remission maintenance in AAV.

Although AAVs were once considered diseases with considerable mortality (80% at 2 years if left untreated), the prognosis has improved significantly over the last 30 years because of improved treatments. Patient survival is now reported to be as high as 45% to 91% at 5 years. Among AAV patients with renal involvement at presentation, 20% develop end-stage renal disease within 5 years.

Henoch-Schönlein Purpura

In mild cases, the therapy for HSP is simply supportive care (i.e., hydration and analgesics). However, glucocorticoids are commonly used to hasten the resolution of symptoms; early use of glucocorticoids has been associated with improved outcomes, especially when there is severe gastrointestinal involvement. In life-threatening cases and in severe acute renal failure, additional immunosuppressive agents or plasmapheresis may be considered. The prognosis of HSP is generally good, with fewer than 1% of patients developing end-stage renal disease.

Medium Vessel Vasculitis

Polyarteritis Nodosa

Treatment of PAN includes glucocorticoids or nonsteroidal anti-inflammatory drugs (NSAIDs) or both. If disease is severe and persistent or relapsing, additional immunosuppressive agents are used, such as cyclophosphamide (especially for gastrointestinal or cardiac involvement), methotrexate, colchicine, or intravenous immunoglobulin (IVIG). In cases of PAN associated with hepatitis B or C, antiviral therapy is required not only for attaining control of the viral infection but also for treatment of the associated vasculitis itself. Corticosteroids and cyclophosphamide have improved patient outcomes, and the 1-year survival rate is now 85%. Prognosis is typically worse with more systemic complications such as renal or neurologic involvement.

Kawasaki Disease

Treatment of Kawasaki disease includes high-dose aspirin (80 to 100 mg/kg/day) for the first 48 hours, then 3 to 5 mg/kg/day. IVIG is standard therapy and has significantly decreased the incidence of coronary artery aneurysm complications in this disease. The initial IVIG dose is 2 g/kg within the first 10 days after presentation, with at least one repeat dose typically given if the first IVIG dose fails to improve the child's condition. The prognosis of Kawasaki disease, if promptly treated, is good; however, approximately 15% to 25% of patients develop coronary artery aneurysms that increase morbidity and mortality.

Large Vessel Vasculitis

Giant Cell Arteritis or Temporal Arteritis

Glucocorticoids are the cornerstone of therapy in GCA. To prevent vision loss, treatment should be instituted immediately (within 24 hours) if clinical suspicion for GCA is high or if visual disturbances are present. The initial dose of glucocorticoids is typically 1 mg/kg/day with a gradual taper. Most patients require a glucocorticoid treatment duration of 1 to 2 years, but it may be longer, especially in those with symptoms of PMR . In PMR without GCA, lower doses of glucocorticoids (10 to 20 mg/day of prednisone equivalent) are effective and provide prompt clinical response.

If patients experience relapse with glucocorticoid tapering, other immunosuppressive agents may be used. Methotrexate was shown in a meta-analysis of three randomized controlled trials to be a beneficial adjunctive agent in reducing risks of first and second relapses in GCA, with a significant decrease in the cumulative dose of glucocorticoids. Low-dose aspirin is an important adjunctive therapy in protecting against cranial ischemic events (level II evidence from two large retrospective studies). Biologic agents in GCA are still under investigation.

Takayasu's Arteritis

Glucocorticoids are also the cornerstone of therapy for TAK; they are typically initiated at a dose of 0.5 to 1 mg/kg/day. Although most patients respond to the initial dose, relapses occur in more than 50% of patients during glucocorticoid tapering. Hence, steroid-sparing agents are often used to aid in maintaining disease remission. The most commonly used steroid-sparing agents are methotrexate and azathioprine. In TAK, unlike in GCA and PMR, the tumor necrosis factor (TNF) inhibitors have shown promise in treating refractory disease. As in GCA, low-dose aspirin is believed to play a beneficial adjunctive role in preventing ischemic complications.

Revascularization interventions are often indicated in patients with TAK whose presenting symptoms include cerebrovascular disease, coronary artery disease, moderate to severe aortic regurgitation, renovascular hypertension, progressive limb claudication, or progressive aneurysm enlargement. Elective intervention should be performed when the disease is quiescent.

In both GCA and TAK, aortitis—a common manifestation of large vessel involvement—can lead to an increased risk of aortic aneurysm and subsequent dissection and rupture. In both GCA and TAK, disease flares occur in most patients, rendering them chronic, progressive and relapsing conditions.

ADDITIONAL CONSIDERATIONS IN TREATMENT

Immunosuppressive therapy is associated with an increased risk of infection. Patients receiving combination therapy with moderate- to high-dose glucocorticoid (>20 mg/day of prednisone equivalents) and another immunosuppressive agent should also receive prophylaxis for *Pneumocystis jiroveci* pneumonia (previously known as PCP). Furthermore, infections can often mimic or result in flares of systemic vasculitis. Glucocorticoid therapy should never be discontinued abruptly, even in the setting of infection, because of the risk of adrenal crisis or disease relapse or both. In most cases, other immunosuppressive agents should be discontinued if infection is suspected or diagnosed.

Glucocorticoid therapy is a common cause of bone loss (osteopenia, osteoporosis). Because significant bone loss can occur even within the first 6 months of therapy, calcium and vitamin D supplementation should be initiated, and a baseline bone density study should be obtained. Consideration should be given to additional bone protection therapies (e.g., bisphosphonates). Methotrexate and cyclophosphamide are teratogenic, and cyclophosphamide may result in premature ovarian failure. These