

# Polymyalgia Rheumatica and Giant Cell Arteritis: Most mayo answers to frequently asked questions

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Polymyalgia rheumatica (PMR) and giant cell arteritis (GCA) are immune mediated diseases of the elderly. Recent studies have revealed that while adequate disease control is seemingly achieved with glucocorticosteroid therapy, both immediate as well as long term complications are not unusual. Patients with GCA are at 17-fold higher risk for thoracic aneurysms, and at 2.4-fold increased risk for abdominal aneurysms over a median of 5 to 6 years of disease. While the overall life expectancy of patients with GCA does not differ from the general population, patients with aneurismal dissection are at markedly increased risk of premature mortality after a median of 2 years.

The causes of these diseases are unknown, although their incidence follows a periodicity consistent with infectious agent exposure, and GCA especially is associated with the shared epitope HLA-BRB104/DRB101. Disease expression may be governed by the pattern of tissue cytokine expression, as patients with GCA and fever have been noted to have low levels of interferon gamma (IF- $\gamma$ ), while

patients with ischemic complications have high expression of IF- $\gamma$  and interleukin (IL) 1- $\beta$ , and patients with large vessel disease have increased IL-2 expression in affected vascular tissue.

Temporal artery biopsy remains the most important and specific diagnostic modality in GCA. While it is preferable to perform the biopsy at initial evaluation, biopsies may remain positive after over one year of glucocorticosteroid therapy. The optimal length of the biopsy specimen is unclear; while a length of about 3 cm is usually recommended, 1 cm may be adequate. Bilateral biopsies are required in about 7% of cases to secure the diagnosis.

Glucocorticosteroids are the only known effective treatment of GCA/PMR. Studies of anticytokine therapies (especially anti tumor necrosis factor) and antimetabolite/chemotherapies to achieve better disease control and alleviate glucocorticosteroid related side effects have been disappointing. Initial treatment of GCA with methylprednisolone 15 mg/kg IV daily for 3 days shows promise as a dose sparing approach.