

**Session Three**

**ADVANCES IN PEMPHIGUS THERAPY**

Dr. P. Régine Mydlarski

9:20 am - 9:40 am  
Saturday, March 24

Pemphigus vulgaris (PV) is a potentially life-threatening autoimmune blistering disease of the skin and mucous membranes. In affected individuals, flaccid bullae develop with the loss of keratinocyte cell-cell adhesion (acantholysis). Immunofluorescent studies reveal IgG antibodies against the keratinocyte cell surface antigens, desmoglein 3 and/or 1. Prior to the advent of corticosteroid therapy, PV was typically fatal. Though disease-related mortality has decreased to 5-10%, the morbidity has risen correspondingly. Current therapeutic options, including prednisone and immunosuppressive agents, are limited by their toxicity profiles. A number of PV patients, unresponsive to high-dose systemic corticosteroids and immunosuppressive agents, have been successfully treated with biologic therapies, plasmapheresis, extracorporeal photopheresis and immunoadsorption. While our treatment options for PV patients are rapidly expanding, uncontrolled clinical trials, case series and anecdotal case reports dominate the dermatologic literature. Randomized controlled trials of mycophenolate mofetil (CellCept), intravenous immunoglobulin (IVIg), infliximab (Remicade), etanercept (Enbrel) and rituximab (Rituxan) are currently underway. The evidence for the use of standard therapies and novel treatments in the management of PV patients will be reviewed.