How is PH in Scleroderma & Connective Tissue Disease Different?
Saturday, June 21: 11:30 a.m. to 12:30 p.m.
Hilton of the Americas: Meeting Room 335 C

Moderators:
Dr. Charles Burger, Medical Director Pulmonary Hypertension Clinic, Mayo Clinic, Jacksonville, Florida
Dr. Kenneth Calamia, Chair, Division of Rheumatology, Mayo Clinic, Jacksonville, Florida
Joy Beckmann RN, MSN, Study Coordinator at Harbor UCLA Medical Center, Torrance, California.

Overview:
Pulmonary Hypertension (PH) is sufficiently challenging even if there are not any associated medical conditions. When associated with scleroderma or other connective tissue diseases, one must also cope with the additional disease. In this session, we will discuss some the challenges of dealing with more than one disease as well as any differences in treatment and expected response. An interactive discussion is encouraged.

Summary:
There are different categories of PH including pulmonary arterial hypertension (PAH) which is further subdivided into idiopathic (unknown cause) and that associated with other medical conditions such as connective tissue disease. The most common connective tissue disease associated with PAH is scleroderma, also called systemic sclerosis. Scleroderma is also divided into categories of limited or diffuse disease. Limited scleroderma which includes calcinosis, Raynaud's phenomenon, esophageal disease, sclerodactyly and telangiectasias (CREST syndrome) may be associated with PAH. More diffuse disease may also produce interstitial lung disease or pulmonary fibrosis (scarring). When patients with scleroderma have been assessed with echocardiography, approximately 13% have PAH. Echocardiography is best viewed as a screening test; therefore, right heart catheterization is often required to confirm PAH. A complete evaluation is often necessary as there are other causes of PH in patients in scleroderma such as pulmonary fibrosis and left-heart disease from chronic systemic hypertension (high blood pressure).
The medications currently approved for PAH can be used with good results in patients with connective tissue disease associated PAH. The most common ones used are the oral medications, at least early in the disease. Occasionally (5 to 10%), patients will respond to calcium channel blockers. The other common oral medications include sildenafil (Revatio or Viagra), bosentan (Tracleer), and ambrisentan (Letairis). More advanced and complicated therapy involves the prostacyclin class of medications. One is inhaled (iloprost) and the other two are continuous infusion therapies (treprostinil or Remodulin and epoprostenol or Flolan). The prognosis of PAH is worse in patients with connective tissue disease even with the most advanced treatment. Unfortunately, the goal for treatment may be primarily to slow the rate of progression of PAH. Although lung transplantation can be considered in patients who do not respond to medical therapies, often the co-existing conditions associated with connective tissue disease disqualify the patient as a potential candidate.