



2009 Fireworks – Pittsburgh, PA

## PIC QUESTION OF THE WEEK: 7/4/11

Q: Please describe the key characteristics and management of adult-onset Still's disease.

A: Adult-onset Still's disease (AOSD) is a systemic inflammatory disorder typically appearing between the ages of 16 and 35. This rare condition may be related to immune dysregulation, genetic mutation, or possess an infectious etiology. Usual signs and symptoms include a spiking fever (generally above 102°F) which is present in approximately 95% of cases (often with sore throat) and a recurring maculopapular rash that is salmon in color and evanescent in nature. The rash typically appears on the limbs and trunk and rarely develops elsewhere. As the disease recurs and progresses, arthritis and arthralgia develop in the knees, wrists, and ankles in nearly all patients. A childhood counterpart of AOSD is known as *systemic* juvenile rheumatoid arthritis (JRA) and formerly referred to as juvenile idiopathic arthritis (JIA) or Still's disease. Although both conditions share common characteristics, *systemic* JRA generally presents between the ages of 3-5, frequently involves other organs (e.g. heart and lungs), and is often less responsive to therapy. The clinical course of AOSD is divided into three phases. The first (*monophasic*) phase is self-limiting and associated with fever and rash. The second (*intermittent*) phase is identified by recurrent *flares* with or without articular involvement and is defined as the polycyclic systemic pattern. Phase three (*chronic* articular pattern) is identified by persistent active disease with significant joint involvement. Diagnosis of AOSD is based primarily on exclusion of other diseases and specific laboratory findings. These include negative rheumatoid factor (RF) and antinuclear antibody (ANA) as well as increased levels of C-reactive protein (CRP) and elevated erythrocyte sedimentation rate (ESR). The key characteristics that differentiate AOSD from rheumatoid arthritis include the absence of RF and ANA as well as increased frequency of carpal and pericarpitate abnormalities. Treatment of AOSD is empirical and often requires combination therapy as the disease progresses. Initial management usually consists of non-steroidal anti-inflammatory drugs (NSAIDs); however, monotherapy with these agents is usually ineffective. Adjunctive corticosteroids such as prednisone and intravenous methylprednisolone may provide substantial benefit. As the disease progresses, addition of methotrexate may be helpful. Immunobiologics such as inhibitors of tumor necrosis factor (TNF), T-cell activation, and interleukin I have been effective in cases of resistant disease. AOSD is not considered curable, but accurate diagnosis and appropriate treatment often result in effective disease management.

### References:

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