



## Department of Internal Medicine TTUHSC - El Paso

### TITLE: CLINICAL PRESENTATION OF SYSTEMIC SCLEROSIS

**Speaker:** Daniel E. Furst, MD, Carl M Pearson Professor of Rheumatology, Department of Medicine/Rheumatology, University of California, Los Angeles, Los Angeles, California

**Introduction:** (e.g. background leading to topic selection)  
Systemic sclerosis is an uncommon disease though we see these patients in our clinic and hospital. Specific visceral involvement may lead to poor prognosis. An update on the evidence based treatment options from a world renowned expert would assist all specialties in managing this disease.

#### Objectives:

- Define the clinical presentation of systemic sclerosis.
- Review potential complications and treatment options of systemic sclerosis.

#### Discussion: Any specific comments regarding the learning objectives

The first learning objective was clearly met by defining the classification of localized and systemic sclerosis. This set the stage for discussing three complications and their treatment options for systemic sclerosis. These were effectively delivered using case-based presentations and challenging the audience to select evidence based diagnostic and treatment options.

#### Conclusions/take home points

Key points for participant to remember

- No internal organ involvement in localized sclerosis.
- Systemic Sclerosis is divided into limited and diffuse sclerosis, which is defined based upon the extent and distribution of skin involvement.
- Systemic sclerosis is a clinical diagnosis. There is a low prevalence of antibodies in scleroderma.
- Prognosis is better in limited scleroderma, unless pulmonary hypertension occurs later in the disease process.
- Interstitial Lung Disease(ILD)
  - o Complete PFT's with DLCO and High resolution chest CT (HRCT) scan are useful tests to diagnose interstitial lung disease and separate interstitial lung disease from pulmonary hypertension as well as to follow the activity/course of interstitial lung disease. HRCT is not good at predicting changes in function, but it rules out other lesions.
  - o Bronchoalveolar lavage has a role in evaluating for lung infection, but its role is controversial for evaluating active lung disease.

- o Once the decision has been made to treat the patient, one option is cyclophosphamide, although the risk-benefit ratio needs to be carefully considered. It is noteworthy that cyclophosphamide significantly improves patient -centered outcomes in ILD
- o Rituximab is a promising drug that may be more effective but is very expensive.

- Skin: Methotrexate, cyclophosphamide, and rituximab improved skin scores.
- Threatened digit is a rheumatologic urgency
  - o Initial management to try calcium channel blocker and add transdermal NTG if needed.
  - o May add phosphodiesterase inhibitor, such as sildenafil, if above measures do not reverse ischemia.
  - o IV prostacyclin infusion is effective.
  - o Oral sildenafil and inhaled iloprost response better in combination.
  - o Bosentan is helpful when ulcers are resistant to healing, and it reduces new digital ulcer formation.
- Scleroderma renal crisis
  - o The most important step is to start an angiotensin- converting enzyme inhibitor and continue to increase dose with the goal to normalize blood pressure.
  - o If blood pressure is inadequately controlled, consider adding a calcium-channel blocker or a diuretic.
  - o Dialyze as needed.
  - o Angiotensin receptor inhibitors are less effective than angiotensin-converting enzyme inhibitors.
- Prognosis can be improved with early detection of complications and initiation of therapy after weighing the risks and benefits carefully with the patient.

### TITLE: END OF LIFE ISSUES IN OLDER ADULTS

**Speaker:** Paul Casner, MD, PhD, Professor of Medicine, Department of Internal Medicine, Texas Tech University Health Sciences Center, Paul L. Foster School of Medicine, El Paso, Texas

**Introduction:** Studies have shown that end of life care in the United States has significant deficiencies. For example, it has been shown that while most patients prefer to die at home the majority of Medicare patients in the United States die in an institutional setting, either in a

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nursing home or an acute care hospital. Furthermore, a substantial proportion of patients die in pain and without adequate emotional support. With the advent of longer life expectancy and an increasing older population end of life care issues will become increasingly important. It is critical therefore, that health care providers be knowledgeable in and prepared to care for older Americans at the end of life.

**Objectives:**

- Identify regional differences in end of life care in the U.S.
- Discuss the appropriate use of opiates in end of life care

**Discussion:** Data from the Dartmouth Atlas Project demonstrates that end of life care varies depending on the region of the country. In regions with higher intensity care such as many urban areas on the east and west coast, patients in the last six months of their lives tend to spend more time in the hospital, more time in the ICU and are less often referred for hospice care. In addition Medicare costs in these regions are much higher than in comparable regions where the intensity of care is less. The reasons for this higher intensity care seem to be based on supply so that a region that has a greater number of beds, greater number of subspecialists and greater number of beds in the ICU will have more intense care at the end of life. However this more aggressive care at the end of life does not often lead to improved patient outcomes nor does it improve patient comfort as studies have shown that these areas of higher intensity care often have worse outcomes and lower patient satisfaction with end of life care. An important aspect of end of life care is appropriate treatment of symptoms. Symptoms can be pain, delirium, dyspnea and problems with feeding. It is important for physicians to be knowledgeable in and familiar with the use of opioids in end of life care. Proper use of opioids requires that patients who have not taken these medications previously should be started on short acting preparations before changing them to long acting opioid preparations. In addition when changing from one opioid to another, equianalgesic dosages must be calculated and because of lack of complete cross tolerance the dosage of the new opioid usually needs to be reduced by 25 to 50%.

**Conclusions/take home points**

- End of life care can be improved by increased utilization of palliative care and hospice care
- Use of feeding tubes in patients with advanced dementia has not been shown to be beneficial either for prolonging life or improving their nutrition or comfort.
- Advanced care planning and usage of advanced directives may be better improved in the future by use of video images rather than verbal descriptions.

**TITLE: INFECTIOUS AGENTS IN CROHN'S DISEASE**

**Speakers:** Rod Chiodini, PhD, Associate Professor, Division of Infectious Disease and William Chamberlin, MD, Associate Professor, Division of Gastroenterology Department of Internal Medicine, Texas Tech University Health Sciences Center, El Paso

**Introduction:** Crohn's disease is a chronic debilitating inflammatory bowel disease of unknown etiology affecting over 500,000 patients in the United States. Recent developments in genetic susceptibility, the intestinal microbiome, and the efficacy of antimicrobial therapy suggest the involvement of an underlying persistent bacterial infection. After 25+ years of debate, gastroenterologists are beginning now to reconsider the role of infectious agents as causative factors in Crohn's disease.

**Objectives:**

- Discuss the possible etiologies of Crohn's disease and the medical controversies associated with the disease.
- Describe the application of PCR and species-specific genes in bacterial identification as related to the complexities of the intestinal microbiome

**Discussion:** At Texas Tech University Health Sciences Center, we have developed a quantitative PCR system to detect the presence of 32 virulence genes and/or pathogenicity islands, representing 16 bacterial species or biotypes, within resected tissues from patients with Crohn's disease. By this approach, we hypothesize that a pattern of detection will develop identifying subpopulation(s) of patients in which bacterial pathogens play a causative role.

**Conclusions/take home points**

- The infectious etiology of Crohn's disease remains a viable etiologic hypothesis
- The leading candidates are Mycobacterium paratuberculosis and/or adherent invasive E. coli.
- Dysbiosis, although clinically important, is not the cause of the disease but rather, the result of the disease.
- Currently used treatments are supportive, do not address the cause of the disease, and are counter-intuitive to the generally accepted concept that patients with Crohn's disease have a deficiency in innate immunity.
- Antibiotics are effective in the treatment of Crohn's disease, but which antibiotics are best is not currently known.

