



Connective Tissue Diseases

East Kootenay Physician Conference

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Disclosures

- Professor Emeritus – University of Calgary
- Funding in Calgary from Pfizer and Amgen –
 - for vaccination education and Rheumatology Yoga program
- Information Technology Officer -Calgary Rheumatology Clinic



Objectives

Connective Tissue Disorders

Understand the relevance of autoantibodies for connective tissue diseases such as Lupus, Scleroderma, Sjogren's , and UCTD (undifferentiated connective tissue disease).

Appreciate key features of clinical history and exam including skin, cardiopulmonary, joint patterns, and neurologic findings.

Learn to identify specific lab abnormalities for hematology, renal and hepatic panels.

Develop an approach to treatment monitoring of steroids, DMARDS, and Biologics in patients with Connective Tissue Disease

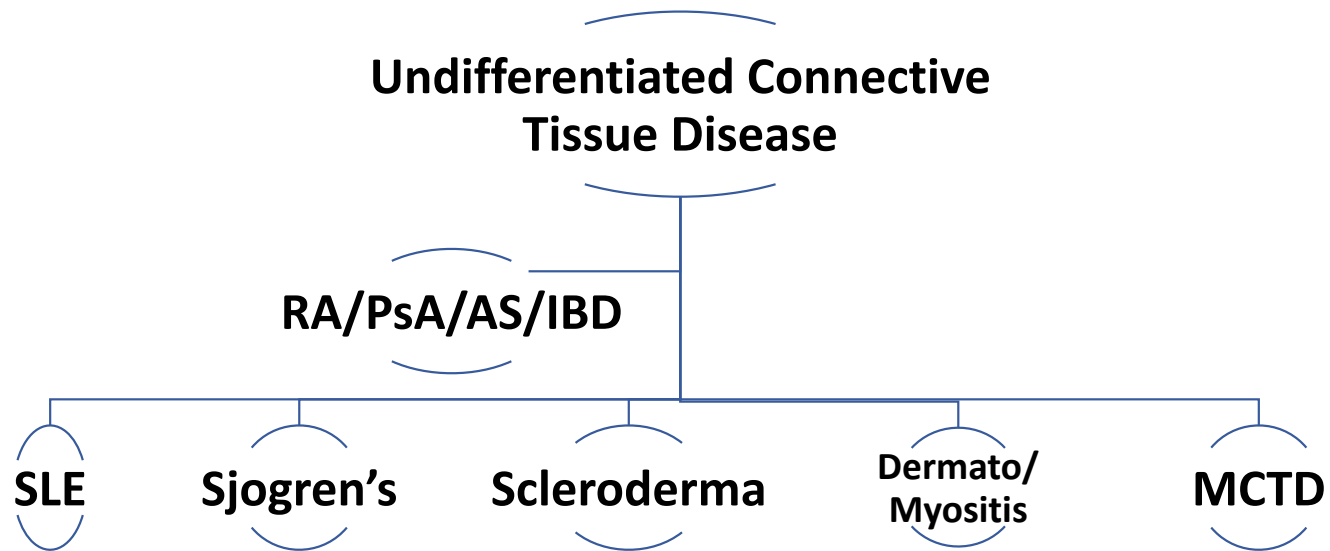


The Information Age (not)

- President Donald Trump once again touted hydroxychloroquine as a treatment for COVID-19, the disease caused by the new coronavirus. This time, the president falsely claimed that “people with lupus” who take hydroxychloroquine “aren’t catching this horrible virus.”



Some general comments on CTD definitions



Case 1 –
25 yo woman with low
energy, general pain, and
“joint aches”

Be thoughtful about ANA
ordering



ANA – Antinuclear Antibodies

ENA – Extractable Nuclear Antigens

- **ANA – antinuclear antibodies testing methodology affects sens/spec**
 - HEP2 IIF (titres 1:80 – 1:2560) – speckled, rim, homogenous, etc
 - Enzyme linked Assay (0.7 and above),
 - Multiplex Immunoassay (Positive/Negative)
- ENA special panels – performed after ANA positive, or strong clinical findings
 - Specific sets available for more definition of:
 - SLE – dsDNA, Smith,
 - Sjogren's – SSA/SSB (Ro/La)
 - Scleroderma – SCL70, Centromere
 - Dermatomyositis - anti-Jo 1



The sensitivity and specificity of ANA has been reported as 40% and 66% (PPV = 29%, NPV = 77%) in a study looking at the diagnosis of any CTD as requested by primary care.

(based on 1994 cohort of patients referred to a rheumatology service in Edmonton Alberta)

Utilization and predictive value of laboratory tests in patients referred to rheumatologists by primary care physicians. J Rheumatol. 1998;25(10):1980-5 Suarez-Almazor M, et al.

- **Key Recommendations** Effective Date: June 1, 2013
- ANA testing need only be ordered once, if found to be positive.
- ANA testing is NOT indicated unless a connective tissue disease (e.g., systemic lupus erythematosus (SLE), scleroderma, Sjogren's syndrome, polymyositis/dermatomyositis) is a significant clinical possibility.
- ANA testing is NOT indicated as a screening test to evaluate fatigue, back pain, or other musculoskeletal pain without other clinical indications.
- ANA testing is NOT indicated to confirm a diagnosis of rheumatoid arthritis (RA) or osteoarthritis (OA).

Statements from BC Ministry of Health “Record of Decision” on Lab Testing – Aug 2021

Testing for ANA is the first step when screening for one of the systemic autoimmune rheumatic diseases (SARD). If the test is negative, the patient is highly unlikely to have one of the SARDs.

ENA testing will not be conducted unless the patient has a positive ANA test within the previous 12 months.

In patients with an ANA titer of 1:320 or higher, ENA testing will automatically be performed as a reflex test.

When the titer for a positive ANA test is 1:80 or 1:160, ENA can still be ordered by a health care provider, but it will not be done automatically.

If ENA is ordered by itself and no ANA test has been performed in the past year, then an ANA test will be run on the patient specimen. The results of the ANA test will determine whether ENA testing is performed.



Case 2–
30 yo
woman with
onset of
Raynauds

Key Features of Clinical History – Fulsome General Systemic Inquiry

Head/Neck – dry eyes/mouth; iritis/scleritis; facial rashes/hair loss

Skin – “rashes”, photosensitivity

Joints – special attention to hands/wrists as well as pattern of presentation

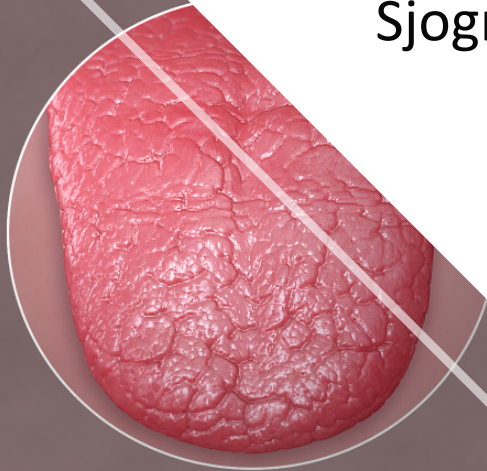
Lungs –SOBOE, persistent cough, chest pain

Cardiovascular – DVT, arrhythmias, pulmonary hypertension

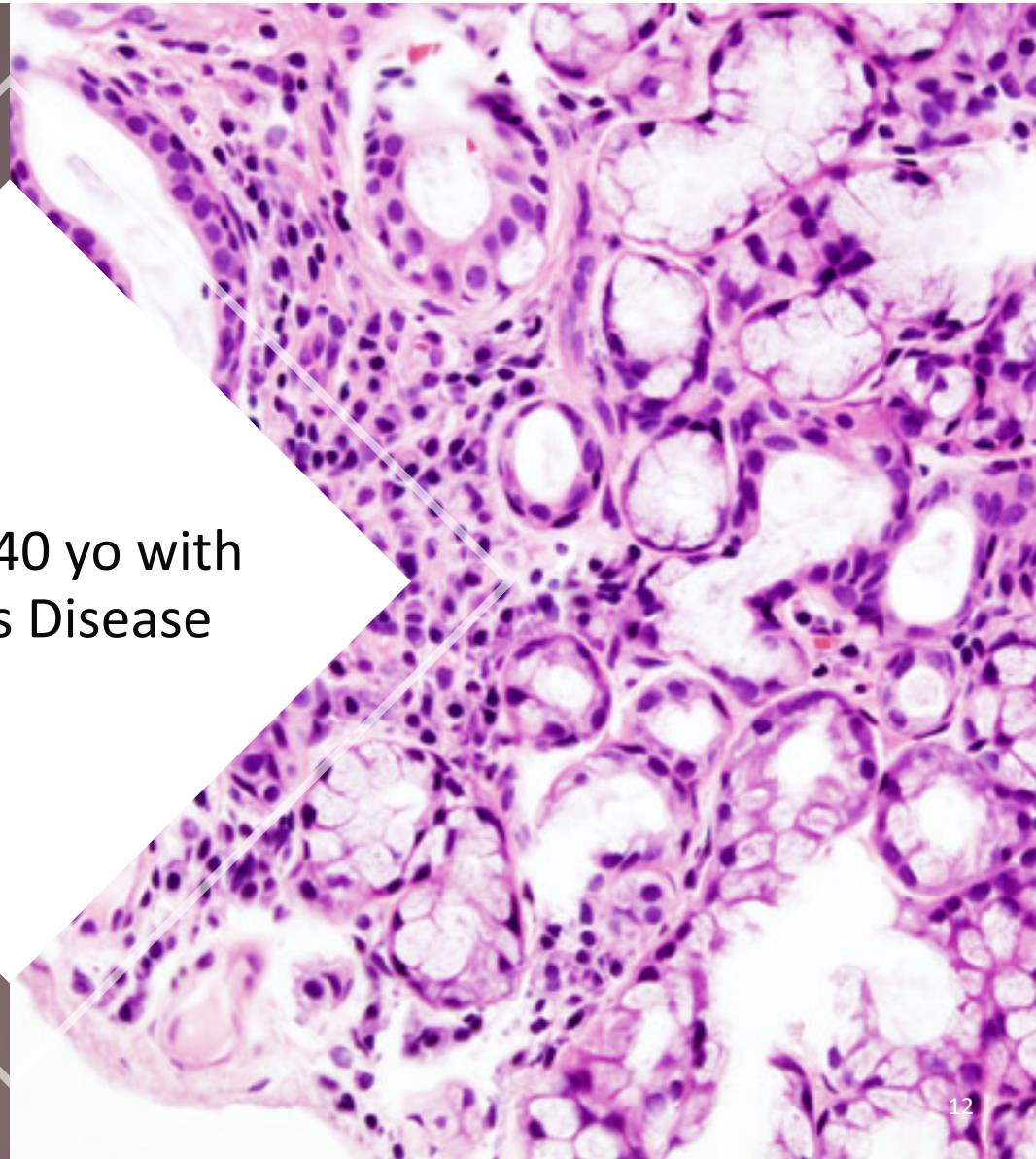
Reproductive – Recurrent pregnancy loss

GI – swallowing difficulties, bowel pattern

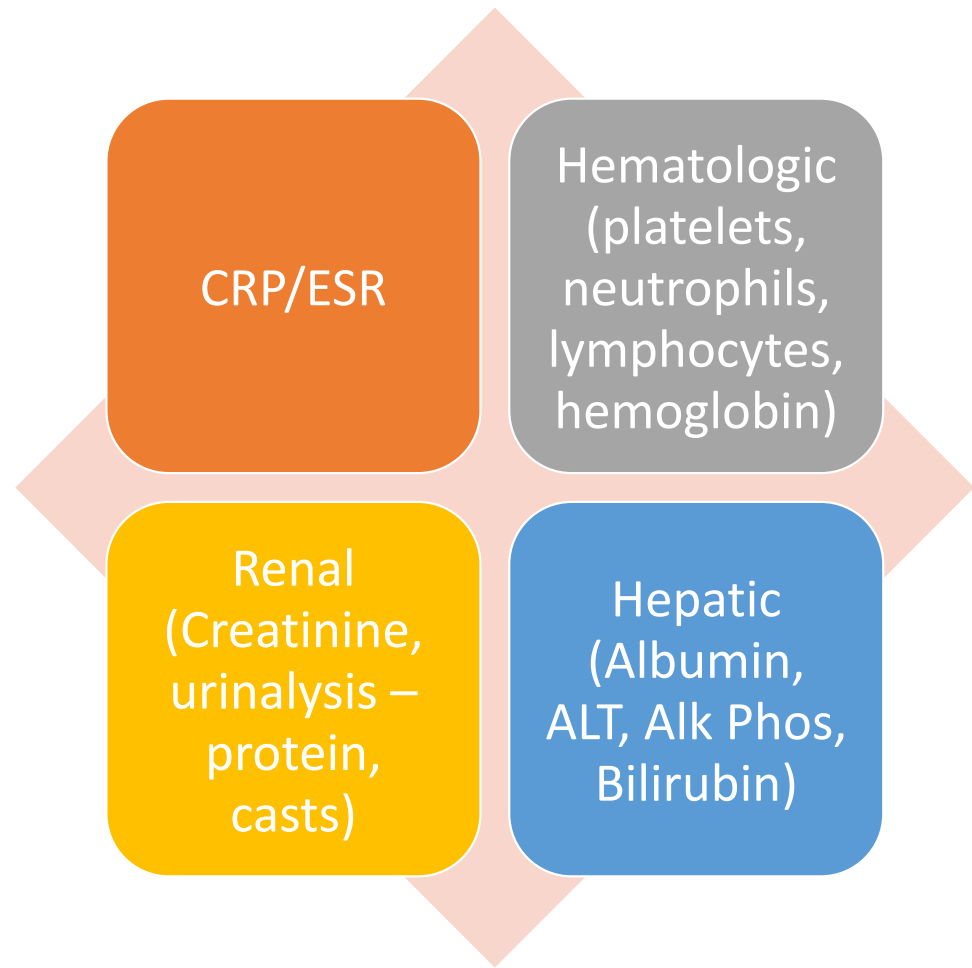
Nervous/muscle system – movement disorder, weakness, sensory loss, TIA/Stroke



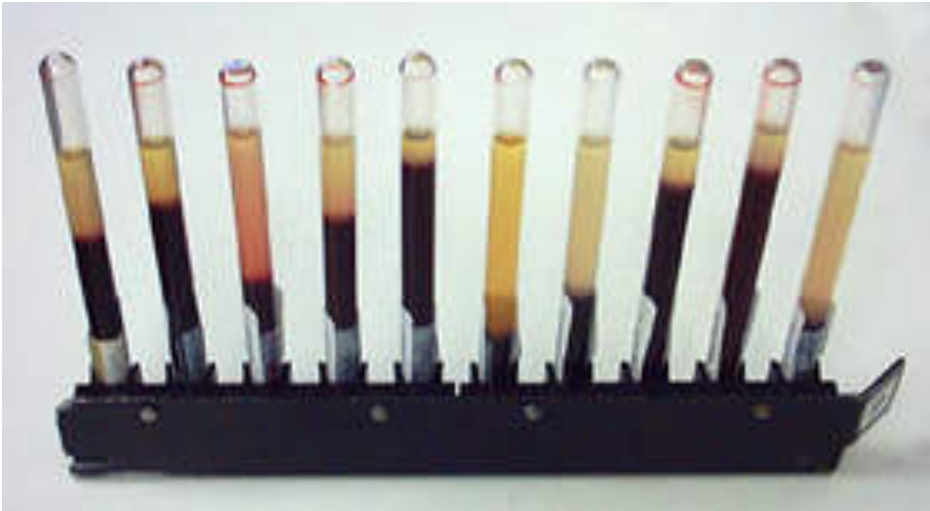
Case 3 – 40 yo with
Sjogrens Disease



Lab Investigations of end organ involvement

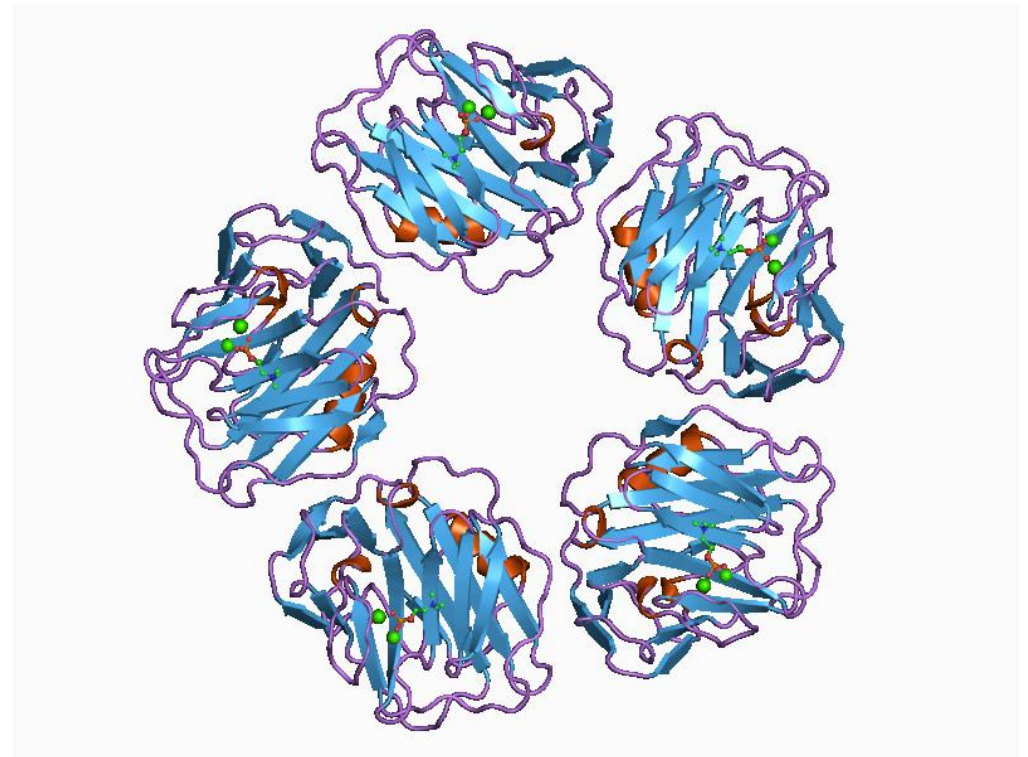


CRP and ESR -



ESR may be reduced in those who are taking [statins](#) and [non-steroidal anti-inflammatory drugs](#) (NSAIDs).

production of [Interferon type I](#) that inhibits the CRP production in liver cells during SLE



Case 4 – 40 yo
Asian male with
SLE



Approach to Therapy

Immunomodulation (not suppression)

Considerations

- Steroids (intravenous, injections, oral, creams)
- Hydroxychloroquine
- Methotrexate
- Azathioprine
- Rituximab

Summary

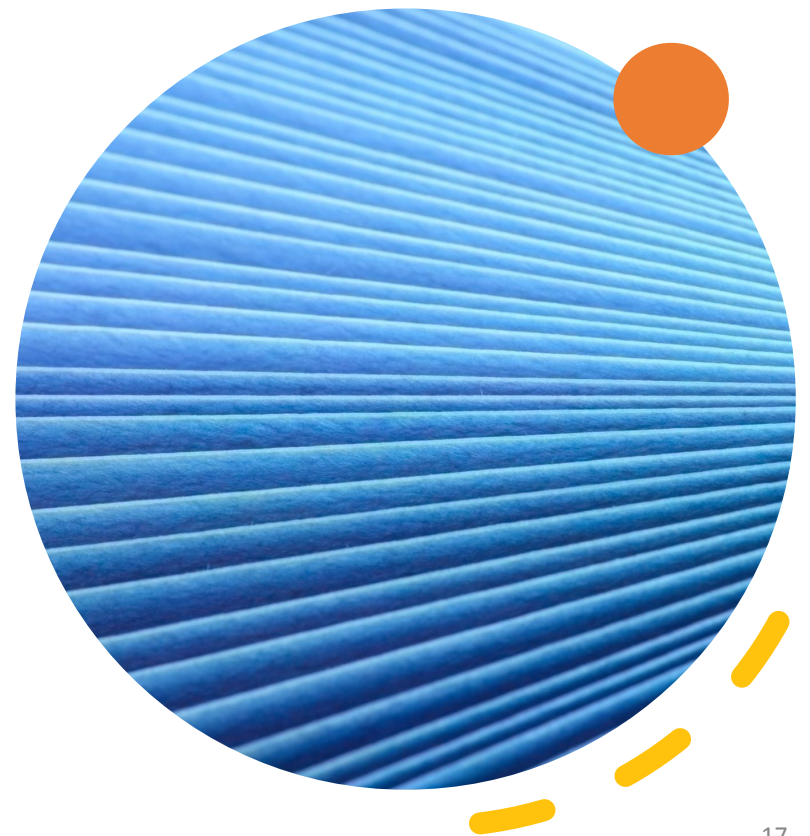
Connective Tissue Diseases are poorly understood by the general public, made more confusing by the Information Age of Internet and Social Media

Autoantibodies (ANA/ENA) are the hallmark of these conditions but also can be fraught with problems of ordering and interpretation

The patient history can be challenging but is essential in determining the likelihood of disease

Examination benefits from a systematic approach from head to toe

Therapy is improving and when applied appropriately can make the difference between life and death



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