

## Answer to Round 3 Quiz 1

Hello everyone!!!

This round has obviously got tougher...Only **SIX** people got the right answer..

Do you like that it has gotten tougher ? If not, (Or even if you do) Please leave your feedback on our social media pages

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Now coming back to the question..

The options available were

- 1) Raynaud's syndrome
- 2) Reynolds Syndrome
- 3) CREST syndrome or limited scleroderma
- 4) Morphea or localized scleroderma

Please see the portions from the question highlighted in yellow, which provided the clues regarding the correct answer

**whitish and bluish discolouration of her fingertips on exposure to cold**  
**tightness of fingers and inability to make a fist**  
**gastro-esophageal reflux symptoms**  
**thickening of the fingers till the MCP joints, Nail fold telangiectasia,**

The whitish and bluish discolouration of her fingertips on exposure to cold is typical of Raynaud's syndrome. (option no 1 ) However that would not explain all her other symptoms and findings. **Hence option no 1 is ruled out**

The tightness of the fingers on history, the Thickening of the skin of the fingers on examination and the nail fold telangiectasia point to secondary raynaud's, most likely scleroderma. (option 3 and option 4) Though scleroderma is classified into localized and systemic, you need to understand that localized scleroderma and systemic scleroderma are 2 entirely different and totally unrelated groups of disease. The only thing common to both groups is thickening of the skin - sclero = thickening , derma = skin.

Localized scleroderma is a dermatological condition with absent or negligible internal organ involvement. It is never associated with Raynaud's **hence option no 4 is ruled out in this patient**

In systemic scleroderma or systemic sclerosis , patients present with Raynaud's followed by thickening of the skin of the fingers which then spreads to other areas of the body in a distal to proximal fashion. If the thickening stops below the elbow or above the knee then that group of patients are said to have limited systemic sclerosis. If it spreads beyond that then they are said to have diffuse variety.

Limited systemic sclerosis patients are more likely than diffuse to have **Cal**cinosis, **E**sophageal dysmotility (contributing to this patient GERD), and **T**elangiectasias (usually on the face). The **CET** along with **R** for Raynauds and **S** for sclerodactyly contributes to the acronym **CREST**. However, generalized itching is not usually seen in CREST syndrome. The hepatosplenomegaly and the liver function abnormalities in addition help to **rule out option number 3**

Let's have a look at her other features which provide a clue

**severe itching**

**increased fatigue**

**total bilirubin is 0.9, conjugated bilirubin is 0.2, SGOT - 57 IU/L, SGPT - 61IU/L, Alkaline phosphatase 387 IU/L**

**Hepatomegaly and splenomegaly.**

**Scratch marks throughout her body.**

Severe itching with scratch marks throughout the body with significantly elevated serum alkaline phosphatase should raise the suspicion of a cholestatic disease. Methotrexate is known to be hepatotoxic. However the dose used in our patient was very small. MTX induced liver problems usually only cause elevation of SGOT, sGPT and not Alk phos.

So once we know that our patient has a cholestatic disease. Let us explore further. In our patient bilirubin is normal, the history is chronic and she has fatigue as well as hepatosplenomegaly. Our patient has CREST which is an autoimmune disease and hence her other problems mentioned above are likely to be autoimmune in nature. The autoimmune disease which can present with the above symptoms is primary biliary cirrhosis (It is now called as primary biliary cholangitis). CREST patients when they have primary biliary cirrhosis in addition (+/- Sjogren's) are said to have **REYNOLD'S SYNDROME. Hence the answer is option 2**

Dr. Reynolds described the condition in 6 female patients in 1970. They had presented just like our patient with pruritus, jaundice and hepatomegaly with marked elevation of serum alkaline phosphatase activity, and had a positive test for serum mitochondrial antibody

Tough luck for this question but be prepared for the next question. Tag your **#smartypants** friend who keeps showing off in class and in front of professors to take up the challenge and become the 1st person to answer the next question on Monday at 6 pm