

PedsCases Podcast Scripts

This is a text version of a podcast from PedsCases.com on “**Physical Examination in a Patient with a Suspected Systemic Rheumatic Disease.**” These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at www.pedcases.com/podcasts.

Physical Examination in a Patient with a Suspected Systemic Rheumatic Disease

Developed by Erin Dockery and Dr. Dax Rumsey for PedsCases.com.

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Introduction:

Hello and welcome to this PedsCases podcast. My name is Erin Dockery and I am a third-year medical student at Western University’s Schulich School of Medicine and Dentistry in Windsor, Ontario, Canada. This podcast was developed with Dr. Dax Rumsey, a pediatric rheumatologist at the Stollery Children’s Hospital and University of Alberta in Edmonton, Alberta, Canada.

Objectives:

The objectives of this podcast are to:

1. List systemic signs and symptoms that may support a rheumatologic diagnosis.
2. Develop an approach to the physical examination of a patient with a suspected systemic rheumatic disease.
3. Demonstrate an approach to narrow a differential diagnosis based on physical examination findings.

Systemic Rheumatic diseases:

Pediatric rheumatology can be a challenging field of pediatrics to learn, as rheumatic conditions are seen less frequently than many other presentations and often present with diverse signs and symptoms. These presentations can range from local to systemic symptoms, from single joint to multi-organ involvement, and from chronic, progressive symptoms to acute presentations. Often, constitutional symptoms, such as fever and fatigue, are also present and, in severe cases, rheumatic conditions may impair growth. Perhaps the most crucial and difficult conditions to diagnose in a timely manner are acute presentations of undifferentiated systemic rheumatic diseases, as these can often be confused with infectious, malignant, or other diseases and notably, can be life-threatening. This podcast will focus on an approach to the physical examination of such patients. Let’s start with a case presentation.

The Case:

You are a clerk completing a rotation on a pediatric ward and are asked to assess a 15-year-old Indigenous female. You note that she has a history of low grade fever and joint pain. You consult a pediatric rheumatologist as you suspect this might be a rheumatic disease. The pediatric rheumatologist would like to know exactly what physical exam

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findings suggest a rheumatic condition and which specific rheumatic disease(s) you have in mind. You head back sheepishly to the bedside to examine the patient. As you can see, fever and joint pain are general symptoms of many conditions and more information is required to narrow your differential diagnosis and to determine whether this patient, indeed, has a rheumatic disease. At this stage, the differential diagnosis is broad and contains both rheumatic and non-rheumatic conditions including, but not limited to:

- Infectious causes, such as septic arthritis or viral arthritis
- Malignancy
- Rheumatic diseases, including:
 - o Rheumatic fever
 - o Systemic juvenile idiopathic arthritis (JIA)
 - o Systemic rheumatic diseases, including systemic lupus erythematosus (SLE), juvenile dermatomyositis, mixed connective tissue disease (MCTD), or scleroderma.
 - o And Vasculitis, of which there are various types (e.g. Kawasaki disease, ANCA vasculitis)

An organized and thorough history and physical examination is required to properly assess this patient and will guide any future investigations. After all, as they say, about 90% of a diagnosis should be made from a complete history and physical examination, with investigations aimed at confirming or ruling out final possibilities. The aim of this podcast is to help you develop a physical exam approach for assessing patients that present with undifferentiated rheumatic disease such as in our patient case. This will help you to narrow your differential and will aid you in providing more informative consultations to your subspecialty colleagues.

Physical exam:

Let's discuss an approach to the physical examination of this patient. Your approach should be broad as patients with systemic rheumatic diseases often present as undifferentiated systemically ill patients. To ensure we don't miss any key information, we have structured our approach from head to toe and by systems. Before even starting the physical exam, think about the demographics of your patient. In this case, an Indigenous adolescent girl is more likely to have a disease like systemic lupus erythematosus (SLE) than, for example, systemic JIA (which is more common in younger patients).

Your physical examination, as always, should start with observations of the patient's overall status and with vital signs. Ask yourself "does the patient look acutely ill or relatively well?". Note the patient's blood pressure, heart rate, respiratory rate, and temperature, as this information will inform your immediate clinical decisions. Many patients may require stabilization before further assessment is obtained.

- Although fever can be a sign of many conditions, including infections and malignancy, prolonged high fevers can be a sign of systemic rheumatic diseases. In particular, high spiking and remitting fevers (i.e. quotidian fevers) are

characteristic of systemic JIA. Hypertension may also be present and can be seen in patients with SLE and other diseases with renal involvement.

Next let's move on to examination of the head and neck. Many key features of rheumatic conditions can be elucidated in the head and neck exam.

- Examine the face and scalp for the presence of any rashes. A malar rash, common in SLE, is often described as a butterfly rash as it spreads across both cheeks and over the bridge of the nose, but spares the nasolabial folds. It can sometimes involve the forehead and is well-demarcated. This rash can appear erythematous or raised and is photosensitive, meaning that it may be precipitated, or made worse, by sun exposure. Discoid rashes, also seen in SLE, are hyperkeratotic, well-demarcated papulosquamous lesions commonly found on the scalp, although they may be present anywhere on the body. These too may be photosensitive rashes and can cause scarring, atrophy, and pigmentation changes to the skin and may be associated with alopecia. They are more common in patients of African descent. Psoriatic plaque lesions may be seen on the scalp of patients with psoriatic arthritis. Patients with juvenile dermatomyositis (JDM) may also present with a malar rash. More classically, however, they present with purplish discoloration around the eyes +/- periorbital edema, known as a heliotrope rash.
- Alopecia, or immune-mediated hair-loss, can be seen in rheumatic conditions, such as SLE. To check for hair-loss, perform the *hair-pull test*. This can be done by grasping 50-60 hairs at the root and sliding your index finger and thumb from roots to ends, collecting any hairs that may have been shed during this process. If several hairs are easily extracted, alopecia may be present. Also, look for signs of thinning and bald patches.
- Next, examine the eyes and mouth for signs of dryness, which is the physical consequence of SICCA symptoms. This can be seen in various systemic rheumatic diseases, such as Sjogren's syndrome and SLE. Upon examination, dry eyes may appear red and irritated. Be sure to check for the presence of cavities, as these are a sign of severely dry mouth. Next, check the mouth and nose for ulcers. These are commonly found on the palate of the mouth and the septum of the nose and can be painless (in the case of lupus). So, the patient or family may not be aware that they are present. While examining the mouth, also check for mucous membrane changes, such as dry, cracked lips and for strawberry tongue, as these may be signs of a vasculitis, particularly Kawasaki disease.
- Lastly, for the head and neck exam, be sure to perform a full lymph node exam as lymphadenopathy may be present in an infectious process, a malignancy, and in various rheumatologic conditions.

Next in our head to toe approach is the chest examination, including complete respiratory and cardiovascular examinations. In this section, we will review some

specific findings that you should keep in mind when examining a patient with a suspected rheumatic condition.

- When completing the respiratory examination, look, listen, and feel for signs of pleuritis, such as decreased breath sounds, dullness to percussion, pleural friction rubs, and pleuritic chest pain with deep breaths. On cardiac exam, the pathognomonic sign of pericarditis is a pericardial friction rub. There may also be muffled heart sounds. Serositis (which includes pleuritis and/or pericarditis) can be seen in various systemic rheumatic diseases such as SLE and scleroderma and in systemic JIA. When completing the cardiovascular examination, it is also important to listen for heart murmurs and extra heart sounds as these may be present in rheumatic fever. Pulses may also be abnormal or absent in certain forms of vasculitis, such as Takayasu arteritis. The Allen test, a special test during which the blood supply to the hand via the radial and ulnar arteries is assessed, can be performed in patients with suspected Raynaud's or other vasculopathies and/or vasculitides affecting the hands. It is also important to check for peripheral edema, as this can be present in various systemic rheumatic diseases, especially when there is renal disease present.

Now that we have gone through some cardiovascular and respiratory findings that can be seen in rheumatic patients, let's move on to the abdominal exam.

- Often rheumatic patients can be tender to palpation of the abdomen. This could represent peritonitis in the acute stage, which is part of the presentation of certain systemic rheumatic diseases, such as systemic JIA. Further, hepatosplenomegaly may be found on examination, notably in patients with systemic JIA or certain other rheumatic diseases.

Next let's move on to examination of the hands. There are many signs of rheumatic disease that can be found when examining the hands.

- Examine the fingers for signs of Raynaud's phenomenon, as this can be associated with multiple rheumatic conditions, such as SLE, scleroderma, and Mixed Connective Tissue Disease (MCTD). Vascular changes in Raynaud's phenomenon impair blood flow to the hands and cause the fingers to change color from white to blue to red, as blood flow returns.
- Rheumatic conditions with vascular involvement may present with ulcers at the tips of the fingers or vasculitic rashes on the hand. Poorly healing cuts on the hands and fingers may indicate poorly controlled Raynaud's, secondary to a rheumatic disease. Diffusely swollen fingers with non-pitting edema can also be seen in MCTD or the early stages of scleroderma.
- Next, exam the nailbeds for any nail changes such as nail pitting, onycholysis, or color changes. If possible, also examine the nailfold capillaries using an ophthalmoscope or other magnifying device. Nailfold capillaries may appear dilated or tortuous in certain rheumatic diseases and there may be fewer capillaries present than expected (called drop out).

As you have probably gathered so far, rashes and skin involvement are common presentations for rheumatic diseases. It is important to thoroughly examine the skin to make sure you do not miss any lesions or rashes that may be present. Let's discuss the key findings of a skin exam for a rheumatologic assessment.

- As mentioned earlier, many and varying rashes are common among rheumatic conditions. Malar or "butterfly" rashes and discoid rashes are characteristically seen in patients with SLE, psoriatic lesions can be seen in patients with psoriatic arthritis and malar rashes, Heliotrope rashes, and Gottron's papules can be seen in patients with juvenile dermatomyositis. Gottron's papules are reddish purple, sometimes scaly, papules that erupt on the extensor surfaces of fingers or other joints. Vasculitic lesions (such as purpuric or ulcerating lesions) can be seen in rheumatic patients if vascular inflammation or damage is present. It is also important to assess skin thickness at different locations on the body (which can be documented using the modified Rodnan skin score), as this can be abnormal in conditions such as scleroderma. Skin in scleroderma may appear thick and tight.

Now that we have completed our examination of the skin, let's move on to the neurological exam and its pertinent rheumatological findings.

- A neurological exam is required to properly assess rheumatic patients.
- Patients with vasculitis may also present with mononeuritis multiplex or peripheral neuropathies in two or more nerve distribution areas. So, it is important to check strength and sensation throughout.
- Systemic rheumatic diseases, such as SLE, may present with exam findings related to cerebrovascular accidents, psychosis, and seizures.
- Abnormal cranial nerve exams may be present in diseases such as sarcoidosis.

Next, a detailed musculoskeletal exam is required to assesses rheumatic patients.

- Many patients with systemic rheumatic diseases will eventually have some form of arthritis, although this may not be present at initial presentation.
- Thus, it is paramount to assess for joint range of motion, tenderness, heat, and effusions in all patients. The pGALS (or pediatric gait, arms, leg, spine) exam is an excellent and commonly used screening tool to assess for joint involvement in pediatric patients. As it is an important component of the rheumatologic assessment, pGALS will likely be the subject of its own future PedsCases podcast and will not be discussed in detail here.
- It is important to assess strength as myositis will present in proximal weakness pattern. This is commonly seen in juvenile dermatomyositis and other idiopathic inflammatory myopathies, as well as occasionally in SLE and overlap disorders.

Lastly, as many rheumatic conditions may be accompanied by vascular changes, it is important to check for localized swelling and tenderness in the calves as this may indicate the presence of DVTs.

Return to the case:

Now that we have reviewed a thorough physical examination approach, let's return to our case.

Upon returning to your patient, you find that she has a low grade fever but otherwise stable vitals. However, she looks unwell. You proceed to inspect her head and neck and find she has a malar rash that spreads across both her cheeks and the bridge of the nose, ulcers on the palate of her mouth, and lymphadenopathy. She also has a discoid rash on her scalp. You complete cardiovascular and respiratory examinations and note that she has a pleural friction rub on auscultation and complains of pleuritic chest pain with deep breaths. On MSK examination, you note she that both her knees are swollen with reduced, tender range of motion. With these distinct findings and the multi-system nature of her presentation, you suspect she has a systemic rheumatic disease, most likely systemic lupus erythematosus (SLE), and you return to the pediatric rheumatologist with your findings. Together, you make a plan.

Take home points:

Now that we have concluded our case study, let's review some key take home points.

1. Systemic rheumatic diseases are autoimmune disorders that can present acutely, with multi-system involvement.
2. The differential diagnosis for systemic rheumatic diseases is broad and includes multiple rheumatic conditions such as lupus, juvenile dermatomyositis, mixed connective tissue disease, and scleroderma, among others, as well as infection, malignancy, and other non-rheumatic autoimmune diseases (e.g. inflammatory bowel disease).
3. Having an organized and thorough approach to the history and physical examination of these patients is crucial. It can help to narrow your differential diagnosis and point you in the right direction in terms of investigations and treatments.
4. When you suspect a systemic rheumatic disease, be sure to contact your friendly local pediatric rheumatologist for advice.

Thanks for listening to this PedsCases podcast!

References:

Woo, P., Laxer, R. M., & Sherry, D. D. (2007). *Pediatric Rheumatology in Clinical Practice*. Springer Science & Business Media.

Cassidy, J. and Petty, R. (2001). *Textbook of pediatric rheumatology*. 4th ed. Philadelphia, Pa., [etc.]: Saunders.