PedsCases Podcast Scripts

This is a text version of a podcast from Pedscases.com on "Henoch-Schlonlein Purpura (HSP)"
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.

# Henoch-Schlonlein Purpura (HSP)

Developed by Vivian G. Szeto and Dr. Rumsey for PedsCases.com.
May 8, 2018

## Introduction:

Hi everyone, my name is Vivian Szeto and I am a medical student at the University of Alberta in Edmonton, Canada. In conjunction with Dr. Rumsey, a Pediatric Rheumatologist at the University of Alberta and Stollery Children's Hospital in Edmonton, Canada, I have created this podcast to provide you with an approach to Henoch – Schlonlein Purpura or simply HSP.

Today's objectives for the HSP PedsCase will include:

1. Define HSP
2. Review the epidemiology and etiology of HSP
3. Recognize the clinical presentation of HSP and potential differential diagnoses
4. Discuss pertinent investigations
5. Discuss management of patients with HSP

In 2012, the revised International Chapel Hill Consensus Conference Nomenclature of Vasculitides changed the official name of HSP to IgA Vasculitis. The name IgA Vasculitis was chosen as a better reflection of the pathophysiology of the disease. Despite this, the disease is still widely referred to by HSP by most practitioners, thus we used the term HSP in this podcast.

## The Case

Let's start with a case! A 9 year old boy presents to the general pediatric community clinic with his mom. You are currently working at this clinic and your task is to take a history. The patient's mother reports that she noticed a rash on her son's legs and buttocks area about a week ago. Her son is now experiencing some abdominal pain along with achy pains in his knees and ankles. He also currently has a low-grade fever. Based on the history and observation of the rash, you suspect this patient may be presenting with HSP. What do you do next?

## HSP Definition

HSP is the most common vasculitis of childhood. It is the result of white blood cells infiltrating vessel walls, resulting in inflammation and necrosis. Children with HSP will present with purpura (with normal platelets), arthritis/arthralgia, and abdominal pain. Complications of HSP include gastrointestinal hemorrhage and glomerulonephritis. The Pediatric Rheumatology European Society (PRES) has proposed the following criteria to diagnose HSP:

Presence of palpable purpura with lower limb predominance, plus at least one of the following 4 features:

i. Diffuse abdominal pain

Developed by Vivian G. Szeto and Dr. Rumsey for PedsCases.com.
May 8, 2018

ii. Biopsy showing typical leukocytoclastic vasculitis or proliferative glomerulonephritis with predominant immunoglobulin A (IgA) deposition
iii. Arthritis or arthralgia
iv. Renal involvement (presence of hematuria and/or proteinuria)

It is important to note that, although part of the criteria, biopsy (either skin or renal) is rarely required. Rather, HSP is typically a clinical diagnosis.

**Epidemiology and Etiology of HSP**

As mentioned previously, HSP is predominantly a childhood disease. It occurs more commonly in boys than girls at a ratio of 1.5 to 1. As well, it occurs most frequently in children between the ages of 3 and 15 years. Children of Caucasian decent have the highest incidence, while HSP is less common in African American children.

It has been reported that HSP is most prevalent during the winter and spring seasons, which supports the hypothesis that HSP is brought on by an infectious agent. Many reports have implicated Group A β – hemolytic streptococci, Staphylococcus aureus, influenza, parainfluenza, Epstein- Barr virus, adenovirus, parvovirus, mycoplasma, Bartonella henselae, and Helicobater pylori as potential triggers for this disease.

**Clinical Presentation and Differential Diagnosis**

Since we have established the criteria for the diagnosis of HSP, let's review how HSP would typically present in the outpatient setting. The onset of HSP is often acute with non-specific constitutional symptoms such as a low-grade fever or malaise.
The cutaneous involvement will include a palpable purpura that is present on pressure-bearing surfaces such as the lower extremities and buttocks. The lesions can range from small petechiae to large ecchymoses to the more rare hemorrhagic bullae. The rash can progress in colour from red to purple to brown and often appears in groups. The purpura may be preceded by a maculopapular or urticarial rash.

Gastrointestinal manifestations occur in approximately two thirds of children within a week after onset of the rash. GI involvement may include: abdominal pain, bleeding, or (rarely) intussusception.

The most common renal presentation is microscopic hematuria with or without proteinuria. Approximately one third of children with HSP will develop renal disease, but it is serious and potentially life-threatening in just 10%. In most cases, serious renal disease will develop within 4 to 6 weeks of the onset of the rash. However, all patients with HSP should be followed for at least 6 months with periodic urinalysis and blood pressure monitoring. Any abnormality on urine testing should be followed by a serum creatinine and, if necessary, referral to a pediatric nephrologist for further evaluation.

Children with HSP will often also be affected by arthritis or arthralgia. Large joints, such as the knees and ankles, are the most commonly affected. However, other areas like the wrists, elbows, and small joints of the fingers may also be involved. On examination, there is often considerable swelling and tenderness and limited range of motion. The joint disease is self-limiting and non-destructive.

Other manifestations that may accompany HSP include edema of the scrotum, eyes, or hands. Additionally, there can be isolated central nervous system vasculitis, seizures, coma, mental status changes, and pulmonary hemorrhage.

It is always important to keep in mind a list of potential differential diagnoses. In the case of HSP, it must be distinguished from other diagnoses such as infections, hypersensitivity vasculitis, acute poststreptococcal glomerulonephritis, acute hemorrhagic edema of infancy, immune thrombocytopenic purpura (ITP), hemolytic-uremic syndrome (HUS), and disseminated intravascular coagulation (DIC), among other conditions.

**Investigations**

Laboratory findings are mostly unremarkable. The CBC will show a platelet count that is either normal or increased, along with a potential left shift in leukocytes. A stool test may confirm the presence of blood loss due to gastrointestinal involvement. Antinuclear antibody and rheumatoid factors are classically absent (and usually unnecessary to order).

If plain abdominal radiographs are ordered, in a patient with gastrointestinal involvement, they may show dilated loops of bowel in the context of decreased intestinal motility. Ultrasound examination may be indicated in a patient suspected of having intussusception. Classically in HSP, however, the intussusception is ileoileal and requires surgical, rather than air enema, correction. A brain MRI/MRA may be indicated in a patient suspected of having CNS involvement.

**Treatment Approach**

For mild HSP, the treatment is supportive with good hydration, analgesics, and non-steroidal antiinflammatory drugs (NSAIDs). Indications for treatment with corticosteroids include severe GI involvement, confirmed renal involvement, CNS involvement, and scrotal edema in boys.

**Key Take Home Points**

1. HSP can present as a multi-system disease.
2. HSP is more common in children, more specifically young Caucasian boys.
3. HSP is diagnosed based on the presence of a palpable purpura along with one of: abdominal pain, characteristic biopsy, arthritis, or renal involvement.
4. It is important to rule out other potential differential diagnoses
5. Treatment is supportive care in mild HSP and steroids for severe HSP.

Thank you for listening to the podcast on HSP. Stay tuned for more podcasts to come!

**References**


1. Brogan P, Bagga A. Leukocytoclastic Vasculitis. Vol 32. Seventh Edition. Elsevier Inc.; 2011. doi:10.1016/B978-1-4160-6581-4.10031-7.
2. Weiss P. Pediatric Vascultis. Pediatr Clin North Am. 2012;59(2):407-423. doi:10.1016/j.pcl.2012.03.013.Pediatric.



Developed by Vivian G. Szeto and Dr. Rumsey for PedsCases.com.

May 8, 2018