**Henoch-Schonlein Purpura**

**Key Points:**
1. HSP is a vasculitic disease often following a URI, usually affecting children but also seen in adults.
2. The main systems involved are the skin, GI tract, kidneys, and joints.
3. Diagnosis is confirmed by skin or renal biopsy, demonstrating IgA deposition.
4. Most recover fully without intervention, but recurrences are common.

**Epidemiology:** Most common in children with over half less than 5 yrs old. Disease is more severe in older children and adults, especially with regards to renal involvement. Often follows an upper respiratory infection but a recent study suggests that an occult malignancy may be the cause.

**Pathophysiology:** Correlation with recent illness suggests an antigenic precipitant to the ensuing vasculitis. Very similar to IgA nephropathy and will see IgA deposits in skin and kidneys on biopsy. Interestingly, pts with renal involvement also have IgG antibodies against mesangial cells.

**Symptoms:** 4 classic ones
1. **Rash** (96%): purpuric, often palpable, distribution mostly over lower extremities. May be urticarial and cause edema, particularly in kids.
2. **Arthralgias** (61%): knees and ankles are targets. Not a destructive arthritis.
3. **GI problems** (48%): cramping abdominal pain, often with vomiting. Appears about a week or more after the rash begins, although there are cases where GI problems occur without a rash. 25% have GIB and 50% have occult blood loss. On endoscopy you see purpuric lesions +/- edema, ulceration, or bowel spasm.
4. **Kidney disease** (37%): Begins few days to weeks after other symptoms. UA can show proteinuria (mild), RBCs, and cellular casts. Many patients will be asymptomatic, but others can develop nephrotic syndrome. On biopsy can see mesangial proliferation and crescent formation; severity of renal disease is correlated with percent of crescent formation.

Other organs less commonly involved: lungs and CNS.

**Diagnosis:** Suspicion from symptomatology should lead to biopsy of skin or kidneys for confirmation of the diagnosis. Skin biopsy shows leukocytoclastic vasculitis with IgA deposition, as seen by immunofluorescence. Kidney biopsy similarly shows IgA deposition.

**DDX:** The big ones are hypersensitivity vasculitis and cryoglobulinemia. Should also consider Wegener’s, anti-GBM, SLE, and other vasculitic syndromes.

**Treatment:** Most recover on their own without treatment. Dapsone may help articular, GI, and skin disease. In those with renal disease, particularly if it is severe, steroids (methylprednisolone followed by 3 months of oral prednisone at 1mg/kg/d) are often given. Other therapies (usually in conjunction with steroids) that have been tried are cyclophosphamide, azathioprine, plamapheresis, and IVIG; none have been studied in randomized controlled trials. Kidney transplant is a final option, but recurrent disease can occur in the new kidney so one should wait 1-2 years until after the rash disappears to transplant.

**Prognosis:** In children pretty good (over 93%)! Initial recovery occurred in 89% of adults. Unfortunately long-term prognosis in adults with HSP and kidney involvement showed that 11% were dialysis dependent and 13% had severe renal disease. Why is this? Probably due to some damage that occurs in the kidneys at time of disease, whereby the other glomeruli participate in “hyperfiltration” to maintain adequate renal function. Over time, this can lead to fibrosis and glomerular damage….ACE inhibitors might help! One retrospective study showed 15 year survival of only 74% in pts with HSP, with carcinoma of the GI or respiratory tracts as the leading cause of death.
Pregnancy risks: A recent study from Finland followed HSP patients for 24 years and found that 70% of women with documented HSP as children had complications of hypertension and/or proteinuria during or after pregnancy.

Recurrences: 1/3 of patients have a recurrent episode, which usually is milder and within 4 months of the initial presentation. Those with kidney involvement have higher incidences of recurrences. Interestingly, in patients who require a kidney transplant, the risk of recurrence goes up if you receive a living-related donor, just like in IgA nephropathy.

References
UpToDate 10.3