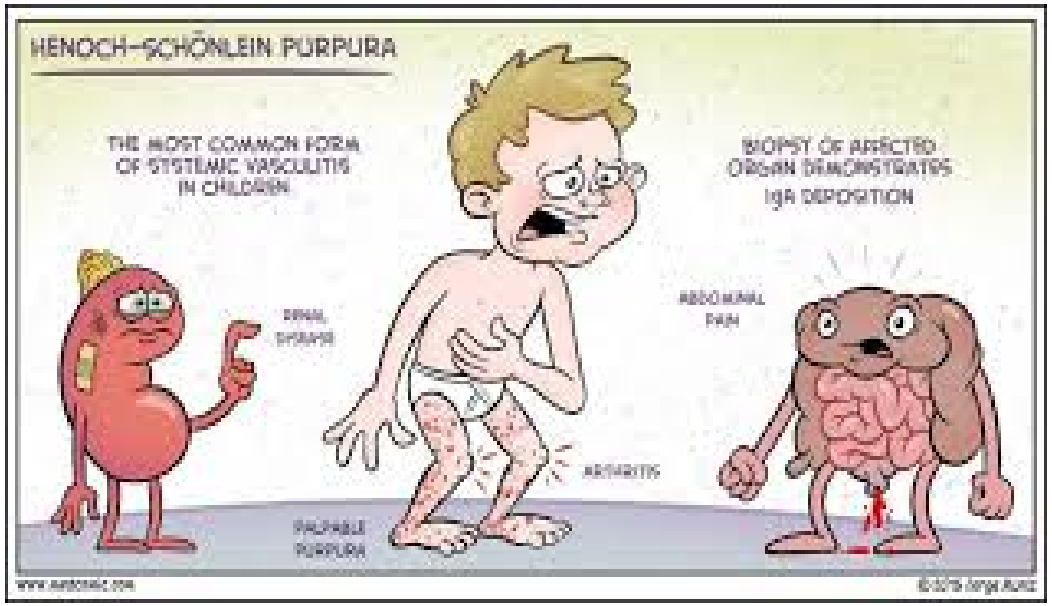
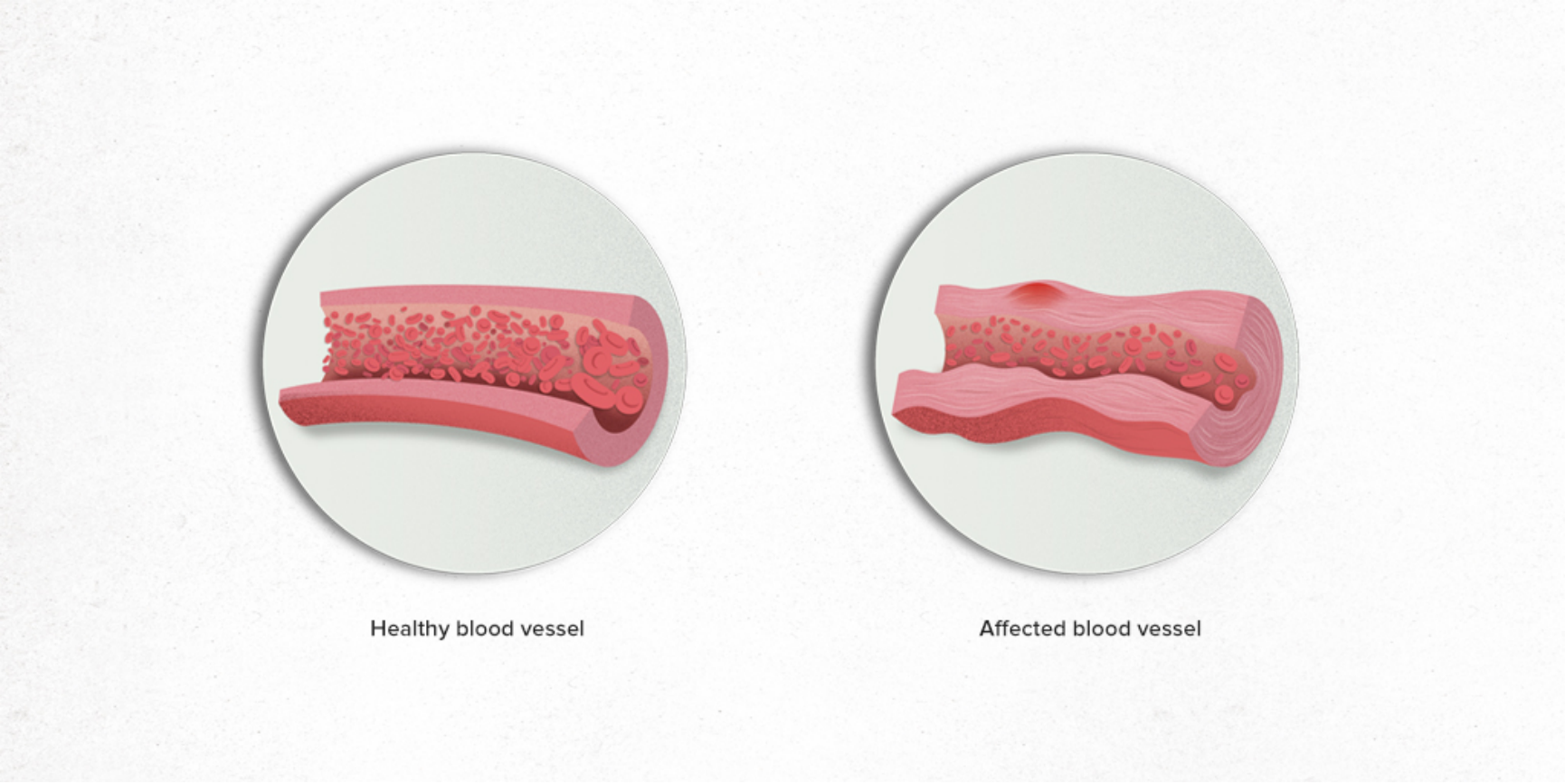
Henoch Schonlein Purpura



Vasculitis



Henoch Schonlein Purpura

IgA vasculitis (IgAV)

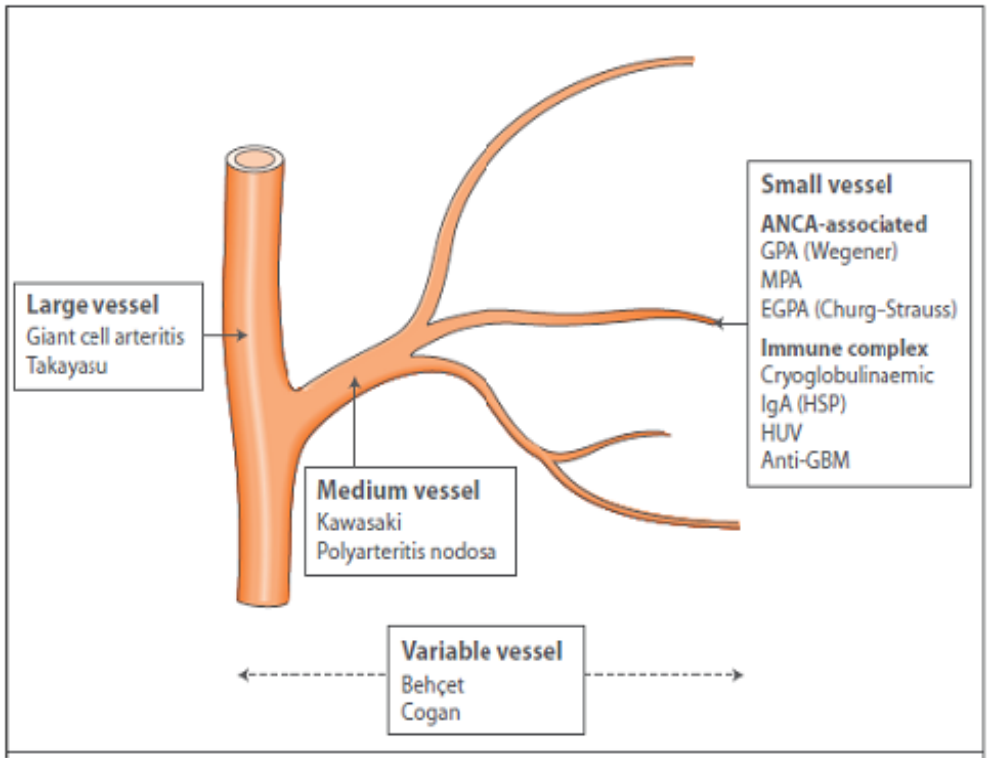
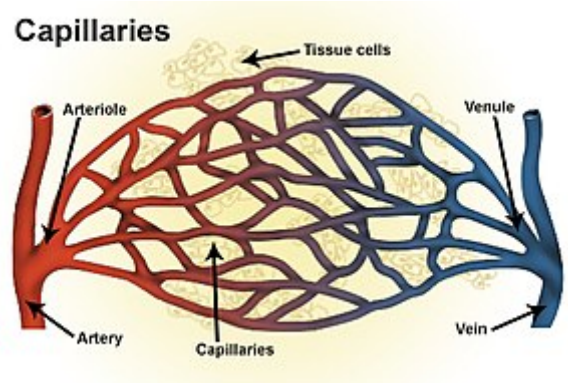


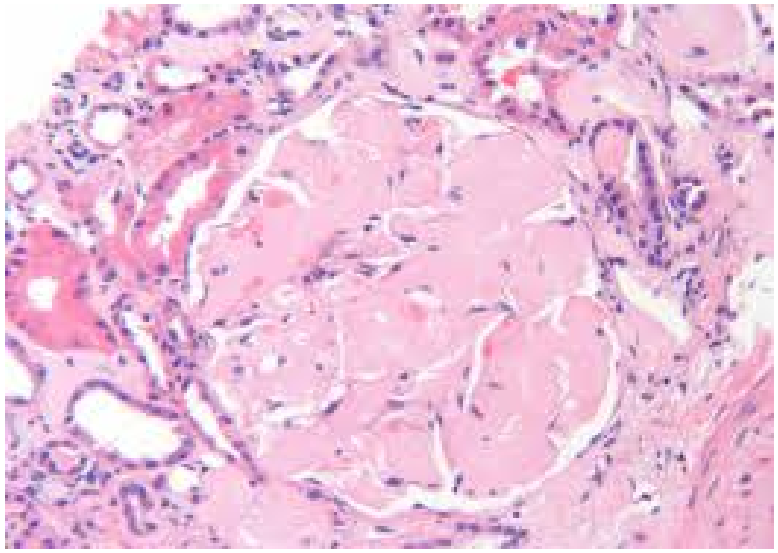
FIGURE 1. Classification scheme for systemic vasculitis.

ANCA antineutrophil cytoplasmic antibody; EGPA eosinophilic granulomatosis with polyangiitis; GBM glomerular basement membrane; GPA granulomatosis with polyangiitis; HSP Henoch-Schönlein purpura; HUV hypocomplementaemic urticarial vasculitis; IgA immunoglobulin A; MPA microscopic polyangiitis



ETIOLOGY

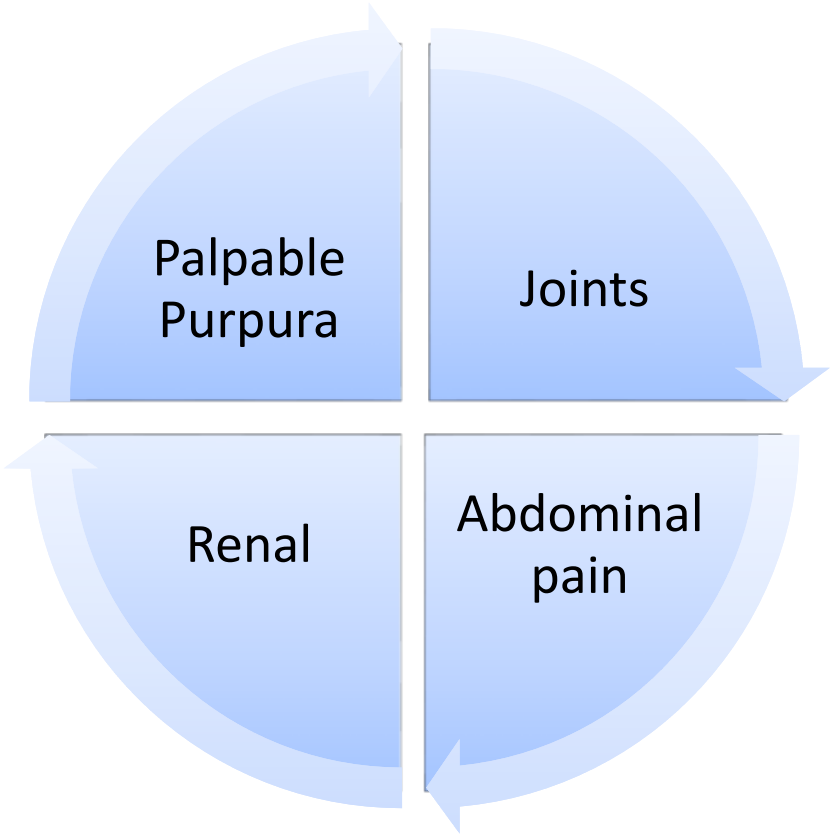
- Henoch-Schönlein purpura (HSP) is a vasculitis of unknown etiology characterized by inflammation of small blood vessels with leukocyte infiltration of tissue.

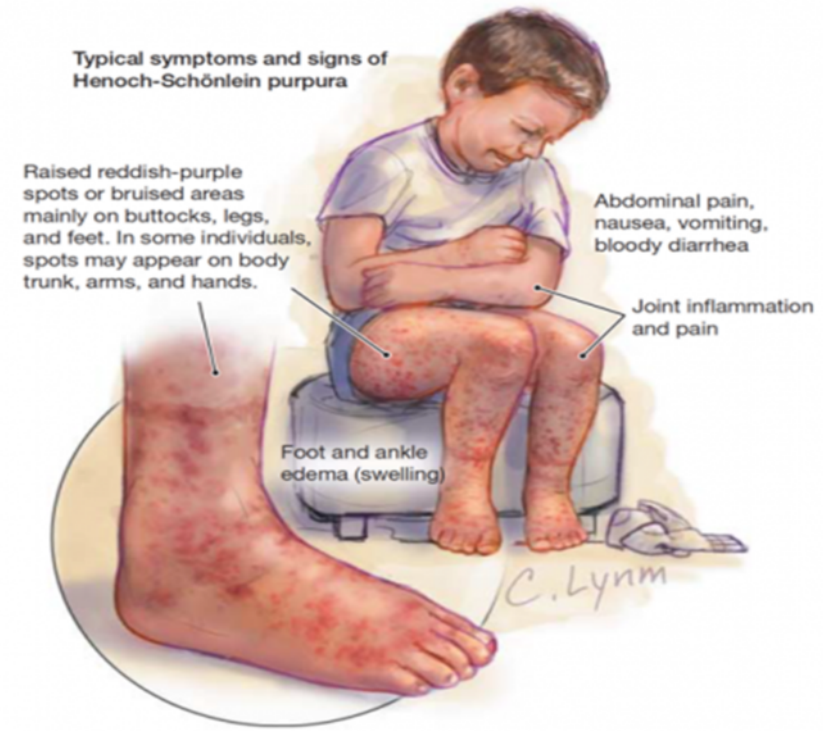


EPIDEMIOLOGY

- HSP is the most common systemic vasculitis
- Children 3 to 15 years of age
- More common in boys
- More frequently in the winter

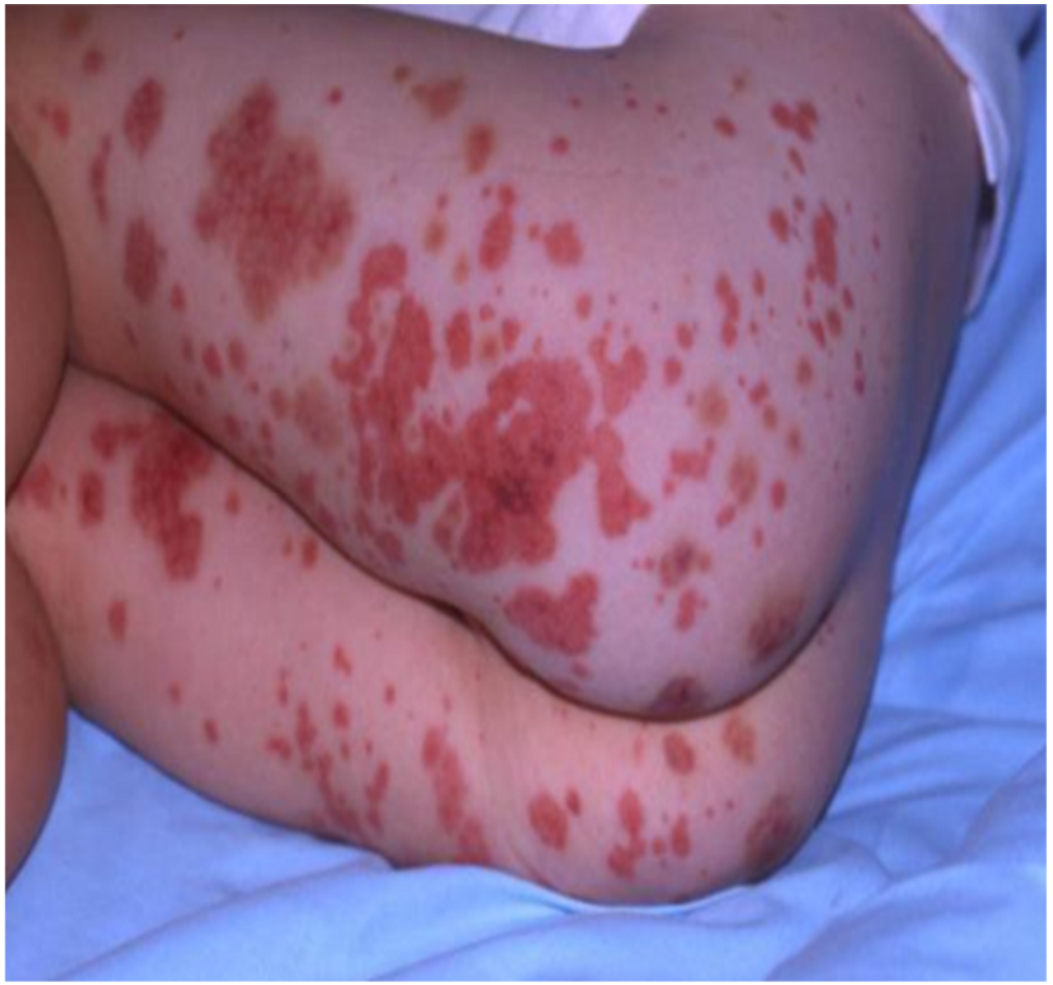
CLINICAL MANIFESTATIONS



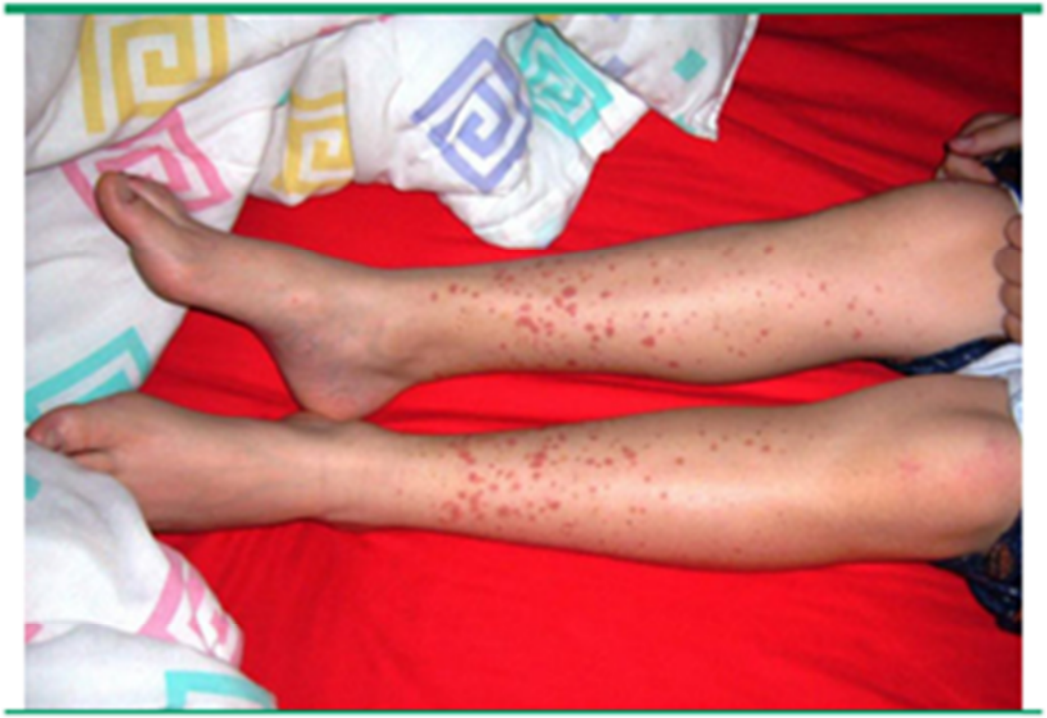


Palpable Purpura:

The hallmark of HSP is palpable purpura , caused by small vessel inflammation in the skin, leading to extravasation of blood into the surrounding tissues, frequently with IgA deposition. The rash is classically found in dependent areas: below the waist, on the buttocks, and lower extremities



Skin manifestations of Henoch-Schönlein purpura (IgA vasculitis)

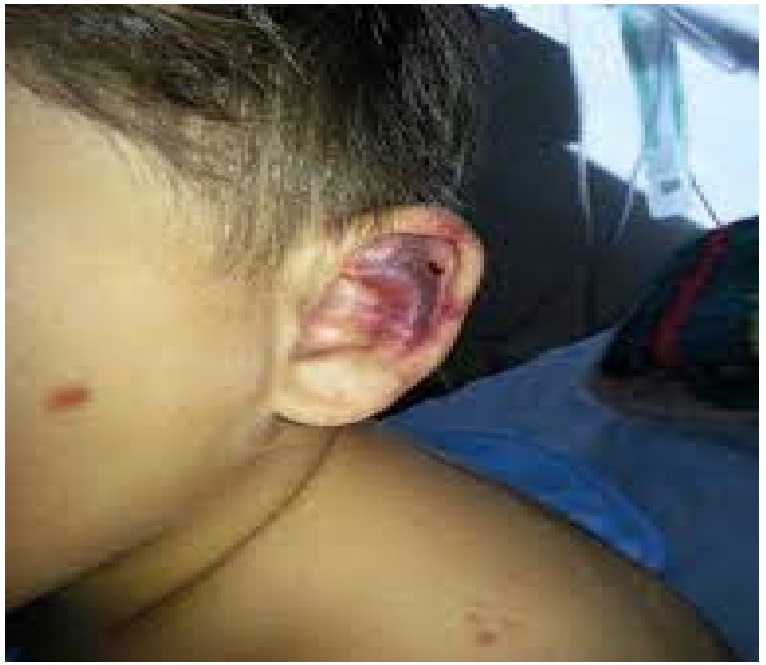


This picture shows the classic skin manifestations of Henoch-Schönlein purpura (IgA vasculitis), with clusters of typical ecchymoses, petechiae, and palpable lesions on the legs in a typical distribution (gravity/pressure-dependent areas).

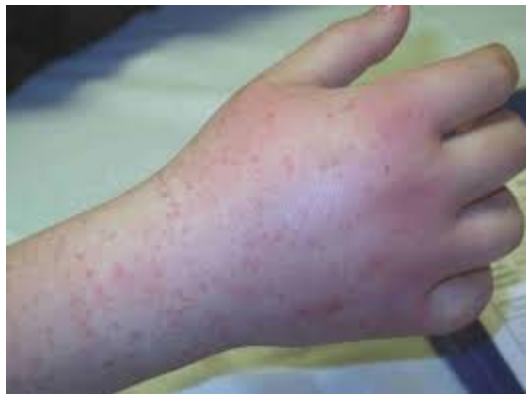
Courtesy of Susan Kim, MD.

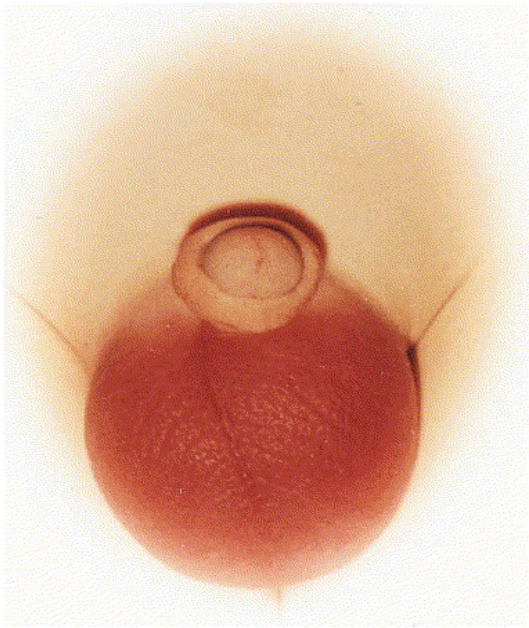
UpToDate®

- The rash can begin as small macules or urticarial lesions but rapidly progresses to purpura with areas of ecchymosis.



- The rash also can be accompanied by **edema**, particularly of the calves and dorsum of the feet, scalp, and scrotum or labia.





Arthralgia/ arthritis:

- 2nd most common presentation 80%

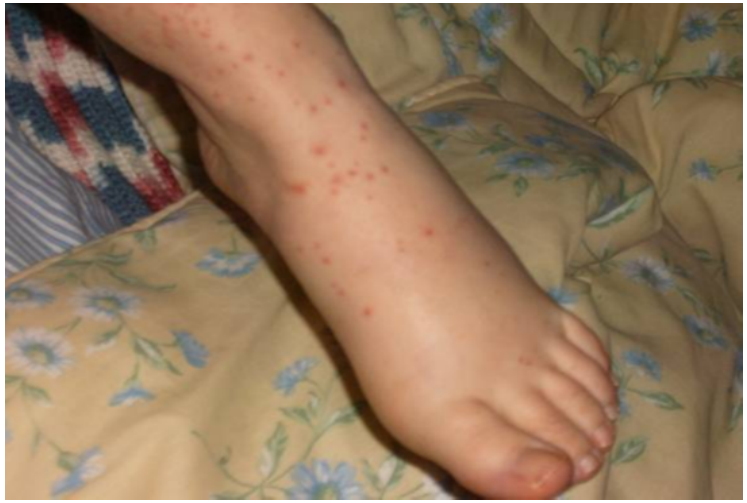
Acute and very painful with refusal to bear weight

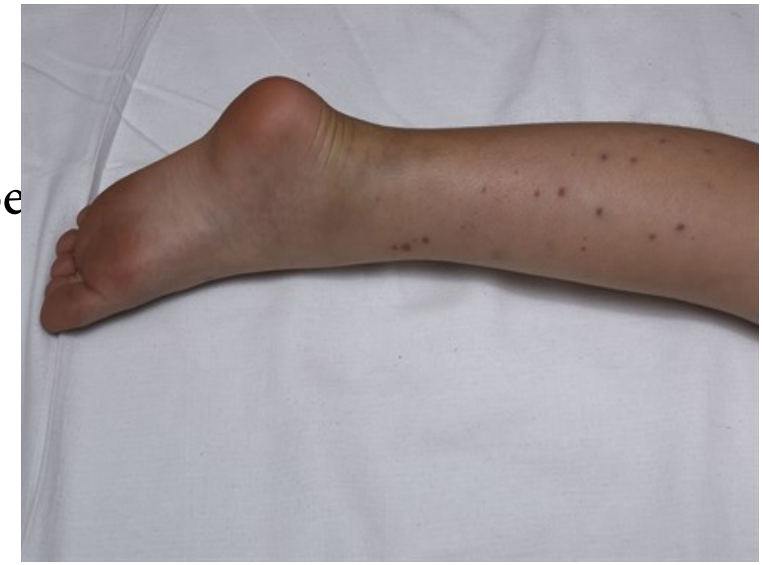
- Oligo articular
- Lower extremity large joints

(ankles and knees)

Does not leave any permanent joint damage; it doe

not typically recur.





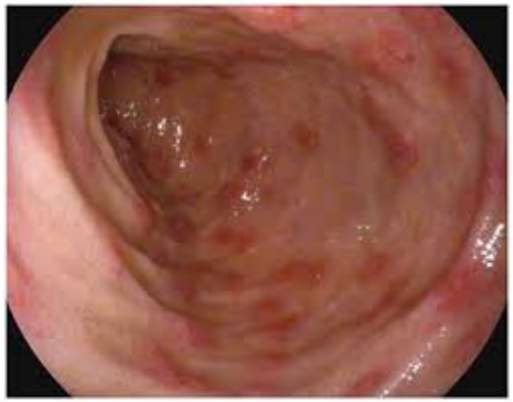
Abdominal Pain:

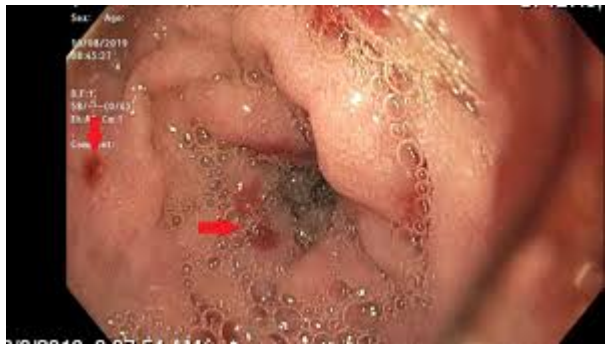
50% of patients complain of colicky pain

During the acute phase of the illness

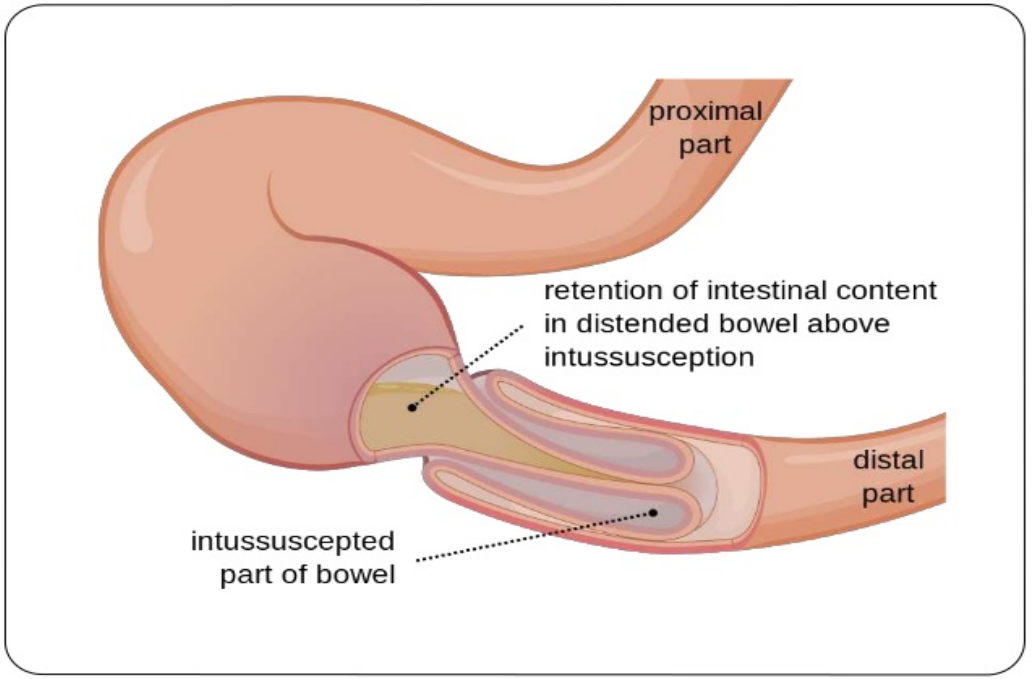
It may precede the onset of rash:

- **Crampy abdominal pain**
- **bloody diarrhea**





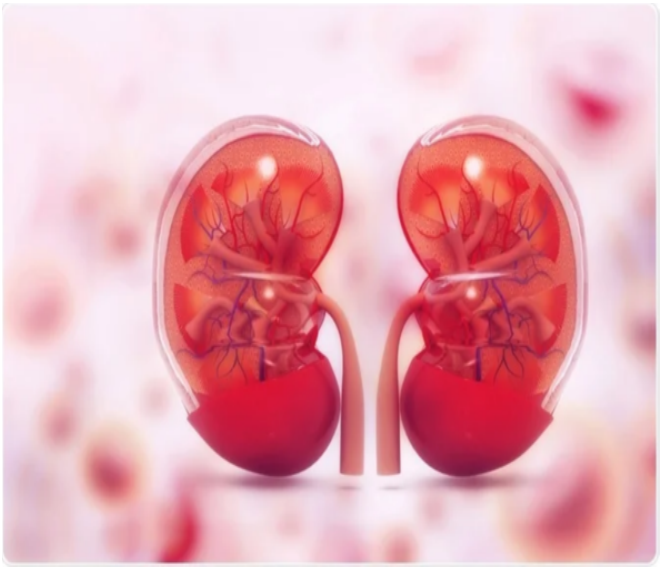
Intussusception



May be followed by complete obstruction or **infarction** with bowel **perforation**.

Renal disease:

- ✓ **One third of children**
- ✓ **Acute glomerulonephritis :**
- ✓ **Hematuria, Hypertension, or Acute renal failure**
- ✓ **Within the first few months of presentation**
- ✓ **Rarely chronic renal disease**



Other organs:

Pulmonary hemorrhage

Encephalopathy

Pancreatitis

Orchitis

Differential diagnosis

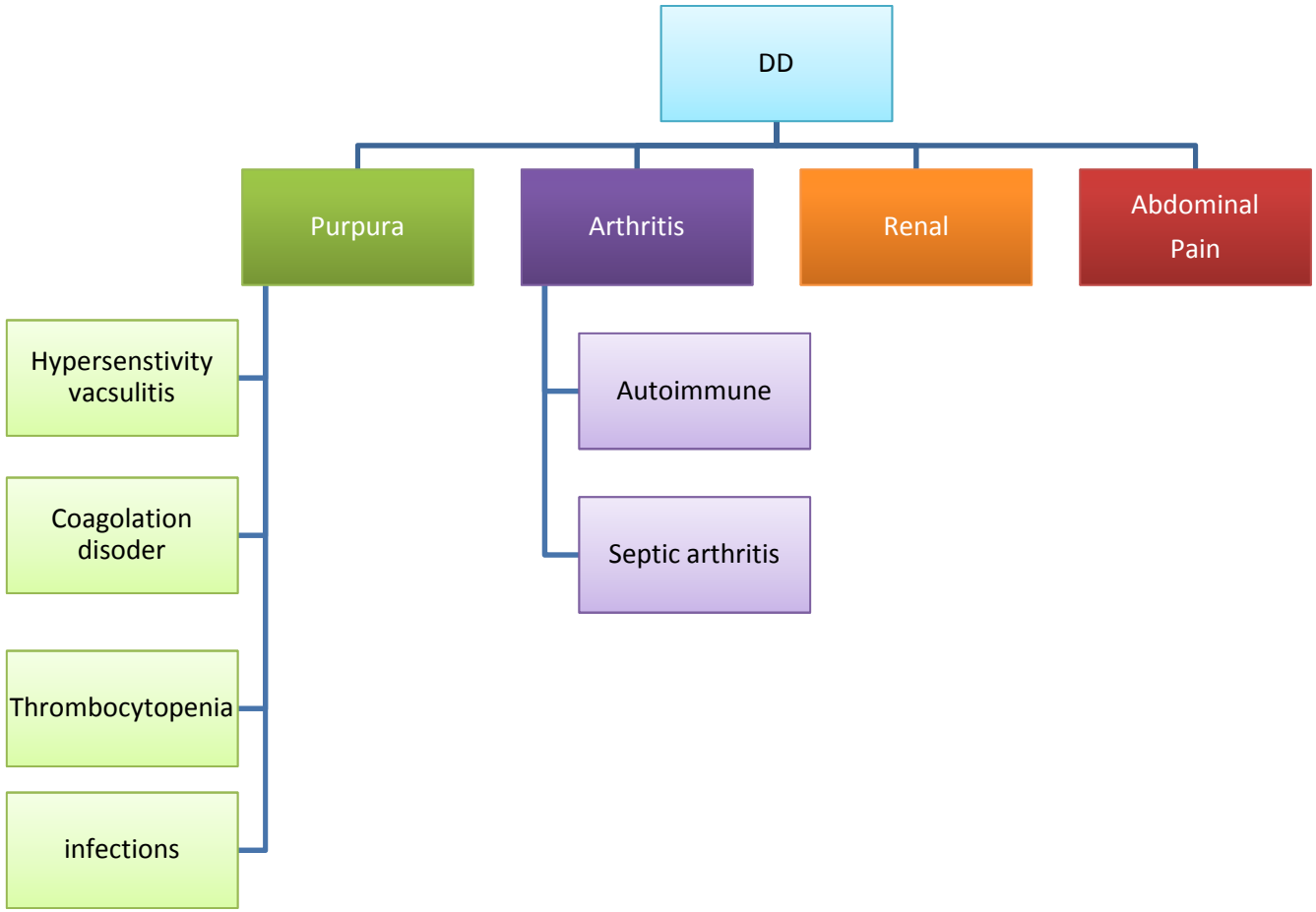


Table 87-1	Criteria for Diagnosis of Henoch-Schönlein Purpura*
CRITERIA	**DEFINITION**
Palpable purpura	Raised, palpable hemorrhagic skin lesions in the absence of thrombocytopenia
Bowel angina	Diffuse abdominal pain or the diagnosis of bowel ischemia
Diagnostic biopsy	Histologic changes showing granulocytes in the walls of arterioles or venules; IgA deposits in vessel wall
Pediatric age group	Age <20 years at onset of symptoms

*The diagnosis of Henoch-Schönlein purpura is based on the presence of two of four criteria.

87.1% sensitivity and 87.7% specificity

Criterion	Description
Mandatory criterion	Purpura or petechiae with lower limb predominance
Minimum 1 out of 4 criteria	1. Diffuse abdominal pain with acute onset 2. Histopathology showing leukocytoclastic vasculitis or proliferative glomerulonephritis, with predominant immunoglobulin A (IgA) deposits 3. Arthritis or arthralgia of acute onset 4. Renal involvement in the form of proteinuria or haematuria

EULAR/PRINTO/PRES: the European League Against Rheumatism, the Paediatric Rheumatology International Trials Organization and the Paediatric Rheumatology European Society (8, 9).

Lab Data

- **CBC Diff :**
- White blood cell count
- **Platelet count** is the most important test **(thrombocytosis)**
- **ESR** 
- **C-reactive protein** 
- Urinalysis screens for evidence of hematuria
- Blood urea nitrogen and creatinine
- Testing the stool for blood

Management

Admission is warranted for the following:

- Sever abdominal pain
- GI bleeding
- Elevated creatinine, HTN, and/ or nephrotic
- Sever joint involvement
- Changes in mental status

Supportive care:

- Adequate hydration, rest and pain relief.

Symptomatic therapy:

NSAIDs:

A short-term course of can be administered for the **acute arthritis**.

Glucocorticoids

- **Gastrointestinal disease**
- **Acute nephritis typically**
- **Orchitis**

(Sever extrarenal complication), Renal involvement

- A typical dosing regimen is prednisone:
- 1 mg/kg/day for 1 to 2 weeks, followed by a taper schedule.

COMPLICATIONS

- Most cases of HSP are monophasic, lasting 3 to 4 weeks and resolving completely.

- The rash can wax and wane, however, for 1 year after HSP.

- Renal involvement rarely may lead to renal failure.

PROGNOSIS

- The prognosis of HSP is excellent.
- Most children have complete resolution of the illness without any significant sequelae.
- There is a long-term risk of progression to
- end-stage renal disease in less than 1% of children with HSP.
- The rare patients who develop end-stage renal disease may require renal transplantation.
- HSP may recur in the transplanted kidney.