Purpura and Vasculitis
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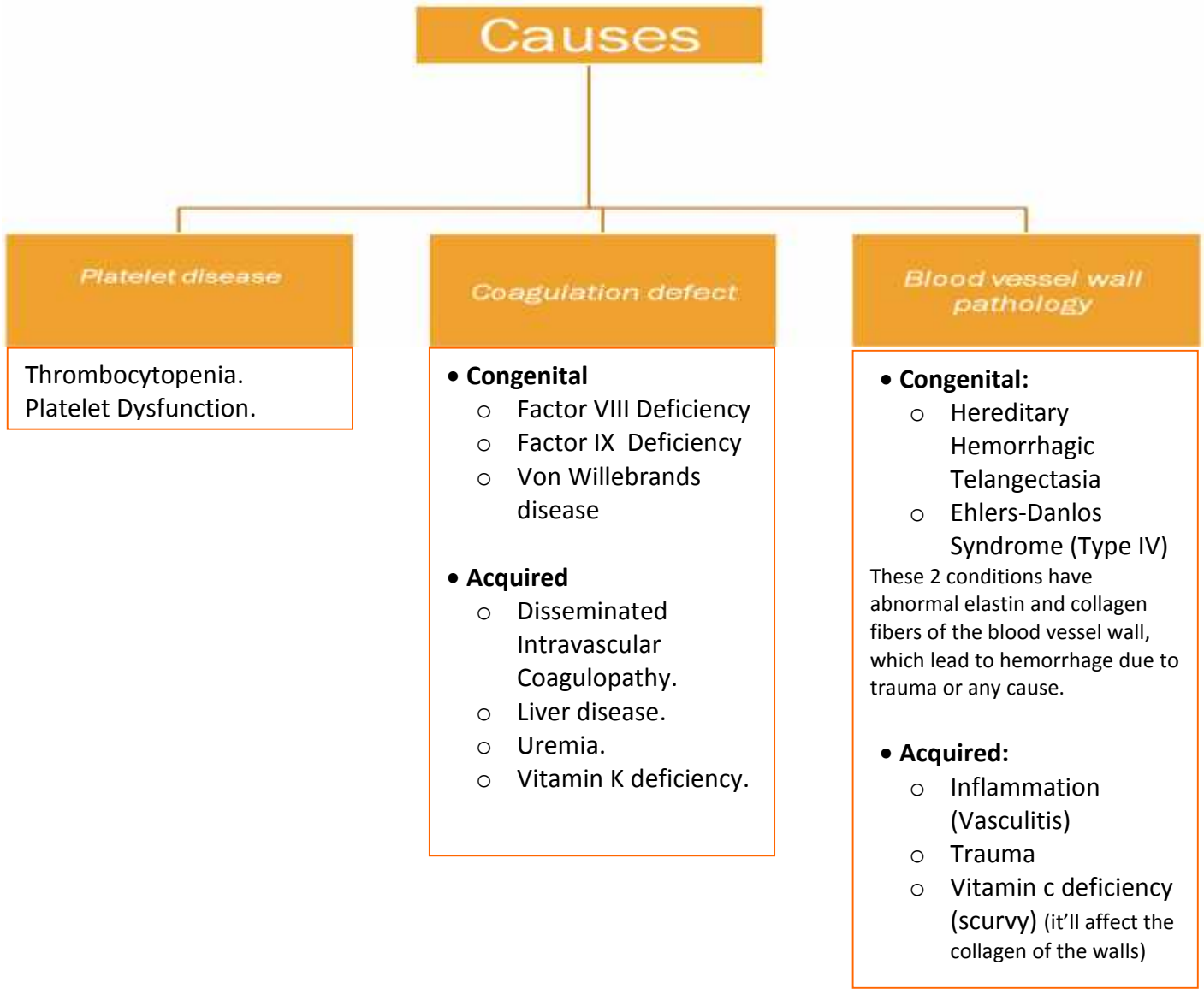
Purpura:

Definition:

Visible hemorrhage into the skin or mucous membrane subdivided as a follow:

- **Petechiae** less than or equal 4 mm.
- **Purpura** (>4mm - < 1cm), which can be either Palpable or non-palpable (macular purpura).
- **Ecchymoses** > or equal to 1 cm.



VASCULITIS:

Definition: A clinic pathologic process characterized by inflammatory destruction of blood vessels that result in occlusion or destruction of the vessel and ischemia of the tissues supplied by that vessel.

Classification:

Table 76.2 Chapel Hill consensus classification

CHAPEL HILL CONSENSUS CLASSIFICATION
Large-vessel vasculitis
• Giant cell arteritis • Takayasu's arteritis
Medium-vessel vasculitis
• Classic polyarteritis nodosa • Kawasaki disease
Small-vessel vasculitis
• Wegener's granulomatosis • Churg–Strauss syndrome • Microscopic polyangiitis (polyarteritis) • Henoch–Schönlein purpura • Essential cryoglobulinemia • Cutaneous leukocytoclastic vasculitis

- **Large-vessel vasculitis:**
  - Aorta and the great vessels (subclavian, carotid).
  - Claudication, blindness, stroke.
- **Medium-vessel vasculitis:**
  - Arteries with muscular wall. E.g. Renal artery, coronary artery.
  - Mononeuritis multiplex (wrist/foot drop), mesenteric ischemia, cutaneous ulcers.
- **Small-vessel vasculitis :**
  - **Capillaries**, **arterioles**, **venules**.
  - Palpable purpura, glomerulonephritis, pulmonary hemorrhage.

Cutaneous Small Vessel Vasculitis (Leukocytoclastic Vasculitis):

Is the most common type of vasculitis and it primarily affecting **post-capillary venules**.

Pathogenesis:

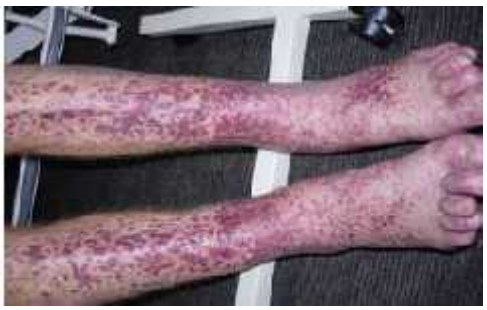
- Many forms of small-vessel vasculitis are felt to be caused by circulating immune complexes.
- These lodge in vessel walls and activate compliment.

Endogens the cause vasculitis:

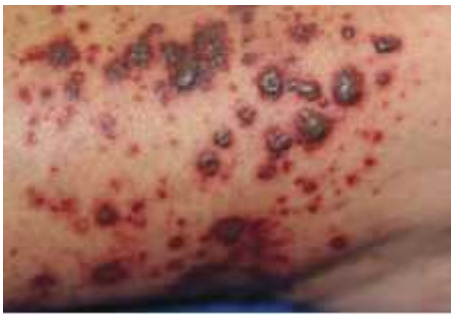
Table 3. Causes of cutaneous vasculitis[5,6]

Infections	
Bacterial	• Streptococcal, meningococcal, urinary tract infections
Viral	• Hepatitis B and C, HIV
Mycobacterial	• Tuberculosis
Connective tissue disorders	• SLE and related conditions • Rheumatoid arthritis • Systemic sclerosis, Sjogren syndrome • Dermatomyositis • Medium vessel vasculitides (Wegener granulomatosis, polyarteritis nodosa, Churg-Strauss syndrome)
Malignancy	• Haematologic – myeloproliferative – lymphoma – monoclonal gammopathy – multiple myeloma
Drugs	Including antibiotics, antihypertensives
Idiopathic	Henoch-Schonlein purpura

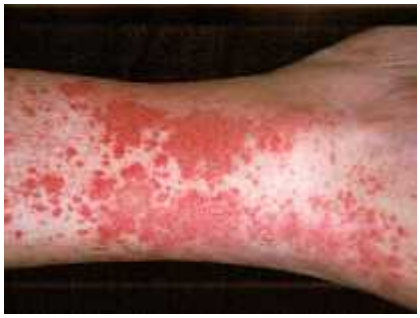
***Streptococcal is the most common.**
***Antibiotic (ABx) is the most common drug.**



- **Palpable purpura** is the hallmark.
- Pinpoint to several centimeters in diameter.
- Early on lesion may not be palpable, Papulonodular, vascular, bullous, pustular or ulcerated forms may develop.
- Predominate on the ankles and lower legs i.e **dependent areas.**



- Mild pruritis, fever, malaise, arthralgia and/or myalgia may occur.
- Self-limiting, typically resolve in 3 to 4 weeks.
- Residual postinflammatory hyperpigmentation may be seen.
- May recur or become chronic, 10% of the patients will have relapses.
- Hemorrhagic vesicles or bullae may develop.

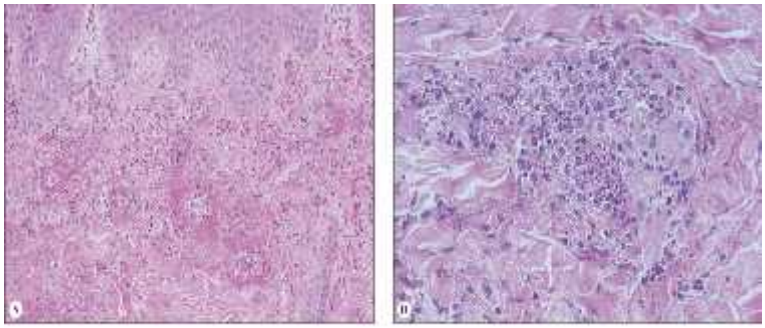


- May be localized to the skin or may manifest in other organs.

- The internal organs affected most commonly include the **joints**, **GIT**, and the **kidneys**.
- Renal involvement present as **glomerulonephritis** (RBC cast).
- The prognosis is **good** in the absence of internal involvement, so we have to check first if there is any internal involvement.

Histology:

Agiocentric segmental inflammation, endothelial cell swelling, fibrinoid necrosis of blood vessel walls and a cellular infiltrate composed of **neutrophil** with **RBC extravasation.**



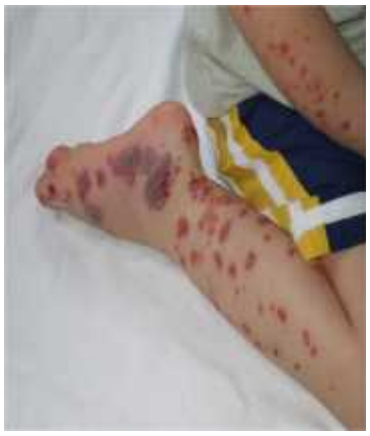
Work-up:

- Detailed history and physical examination.
- History should focus on possible infectious disorders, prior associated diseases, drugs (ABx) ingested, and a thorough review of systems.
- **CBC**, strep throat culture or **ASO titer**, **Hep B & C serologies** and **ANA** are a reasonable initial screen.
- Urinalysis (RBC cast), to rule-out renal involvement. **(very important)**

Treatment:

- Treatment of cause.
- Symptomatic treatment (if skin is only involved): rest, NSAIDS, Antihistamine, Topical steroid.
- Severe visceral involvement may require high doses of **corticosteroids** with or without an **immunosuppressive agent**.
- Immunosuppressive agents for rapidly progressive course and severe systemic involvement.

HENOCH-SCHONLEIN PURPURA (HSP):



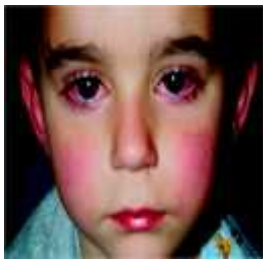
- Primarily occurs in male children.
- Peak age 4-8 years.
- Adults may be affected.
- A viral infection or **streptococcal pharyngitis** is the usual triggering event.
- In about 40 % of the cases the cutaneous manifestations are preceded by systemic symptoms; mild fever, headache, joint symptoms, and abdominal pain for up to 2 weeks.

- **Characterized by intermittent purpura, arthralgia, abdominal pain, and renal disease.(Important)**

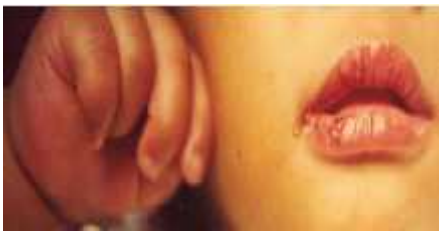
To differentiate HSP from the Cutaneous Small Vessel Vasculitis (Leukocytoclastic Vasculitis):

- Typically purpura appears on the **extensor surfaces** of the extremities.
- Become hemorrhagic within a day and fades in **5 days.**
- New crops appear over a few weeks.
- May be associated with:
  - **Pulmonary hemorrhage. (Important)**
  - Abdominal pain and GI bleeding.
- GI radiographs may show "cobblestone" appearance.
- **Renal manifestations may occur in 25% or more but only 5% end up with ESRD, so you must evaluate each patient for renal involvement. (Important)**
- The long-term prognosis in children with gross hematuria is very good; however, progressive glomerular disease and renal failure may develop in a small percentage.
- **IgA**, C3 and fibrin depositions have been demonstrated in biopsies of both involved and uninvolved skin by immunofluorescence techniques. **(Important)**

MUCOCUTANEOUS LYMPH NODE SYNDROME (KAWASAKI'S DISEASE):



- Medium-vessel vasculitis.
- It affects the skin, mucous membranes, and lymphnodes.
- Predominantly seen in children less than 5 years of age.
- Occurs most often in Japan
- To make the **diagnosis** a patient should have a **fever** above 38.3 C for 5 days plus 4 of the 5 following criteria;
  - Edema of hands and feet, after 2 weeks it will present with exfoliation of the hand and feet.
  - Polymorphous exanthema, can present as urticaria.
  - Nonpurulent bilateral conjunctival injection.
  - Changes in the lips (sharp lips) and oral cavity (strawberry tongue).
  - Acute, nonpurulent cervical adenopathy



- **Coronary arterial disease** occurs and **thrombocythemia** may occur. **(Important)**
- In combination vessel occlusion may occur and the subsequent **MI**, which occur as the child is recovering from the acute illness.

- **Treatment: (Important)**
  - **IVGG** is the cornerstone of treatment
  - Antiplatelet therapy with **aspirin** is recommended.

GOOD LUCK..
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