INDICATIONS FOR TESTING
Unexplained systemic illness with organ involvement including
- Renal
- Pulmonary
- Dermatologic
- Rheumatologic (arthritis, myositis, serositis)
- Neurologic (mononeuritis multiplex)

ORDER
- Urinalysis (UA)
- C-reactive protein (CRP) or sedimentation rate (ESR)
- Imaging
- Biopsy

CRP or ESR elevated
Hematuria

Systemic illness and 2 of the following
- Skin involvement (livedo reticularis, tender subcutaneous nodules)
- Myalgias or muscle tenderness
- Systemic hypertension
- Mono- or polyneuropathy
- Abnormal UA
- Testicular pain or tenderness
- Signs and symptoms suggesting organ involvement vasculitis

AND
- Biopsy demonstrating small and medium size artery vasculitis
OR
- Angiographic abnormalities demonstrating vasculitis (e.g., microaneurysms)

Polyarteritis nodosa

Kawasaki disease

Typical criteria
- Fever of ≥5 days and 4 of the following
  - Erythema of palms and sole with possible desquamation
  - Polymorphous rash
  - Bilateral conjunctivitis, nonpurulent
  - Oral cavity changes including erythema, dryness and cracking of lips, strawberry tongue and diffuse erythema
  - Cervical lymphadenopathy (>1.5 cm diameter)

At least 1 of the following
- Decreased peripheral arterial pulses
- Blood pressure difference >10 mmHg between extremities
- Bruits over aorta or its major branches
- Hypertension
AND
- Arteriography demonstrates stenosis

Takayasu arteritis (Aortic arch syndrome)

ANCA -

Palpable purpura
AND
- At least 1 of the following
  - Diffuse abdominal pain
  - Arthritis or arthralgia
OR
- Biopsy demonstrating IgA deposition (small vessel disease)

IgA vasculitis (formerly Henoch Schönlein Purpura)

ANCA +
(antiproteinase 3 predominates)

Any 3 of the following
- Biopsy demonstrating granulomatous inflammation with small size vasculitis
- Abnormal UA
- Nasal-sinus inflammation
- Subglottic, tracheal or endobronchial stenosis
- Abnormal chest x-ray or chest CT

Granulomatosis with polyangiitis (formerly Wegener granulomatosis)

Note: Other vasculitic syndromes (e.g., microscopic polyangiitis) exist, but are rare in children

Other suggested testing
- Drugs
- Neoplasm
- Infection (HCV, Rickettsia, Lyme)
- Autoimmune

References