INDICATIONS FOR TESTING
Unexplained systemic illness
Multiple system involvement (renal, pulmonary, dermatologic)
AND
The following have been ruled out:
- Infection
- Neoplasm
- Drug effect

To determine systems involved; ORDER
- Urinalysis (UA)
- C-Reactive protein (CRP) or Sedimentation rate (ESR)

CRP or ESR elevated
Hematuria on UA

No
Yes

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)

Kawasaki disease

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)

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 (eg, microaneurysms)

Polyarteritis nodosa

IgA vasculitis
(should have MPO/PR3 antibodies)

ANCA -

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

ANCA -

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

ANCA +
(antiproteinase 3 predominates)

Granulomatosis with polyangiitis
(formerly Wegener granulomatosis)

Notes:
Other vasculitic syndromes (eg, microscopic polyangiitis) exist, but are rare in children
*(American Heart Association, 2004)