Henoch-Schönlein Purpura

JP Thompson PA-C

A Zebra Talk

A diagnosis not entertained is a diagnosis not made.

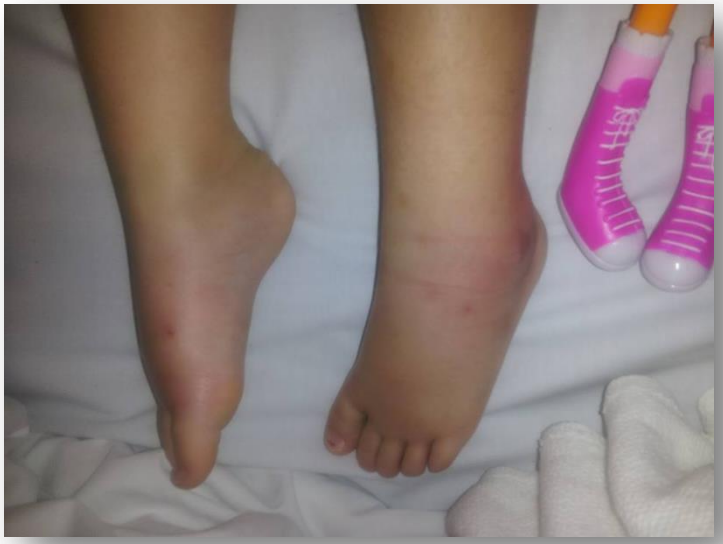
Henoch Schönlein Purpura

Objectives:

- Describe Henoch Schönlein Purpura
- Review treatment options
- Discuss pros and cons of treatment options

What is it?

Henoch–Schönlein purpura (HSP) is one of the most common forms of systemic vasculitis manifested in childhood.



Who named it?

The first case of HSP was described by William Heberden (1710–1801) in 1801. He described a 5-year-old boy with a purpuric **eruption,** macroscopic hematuria, abdominal pain, bloody stools, vomiting, arthralgia, and edema.

HeberdenW.Commentarii Di Morborium Historia et Curatione. London: Payne, 1801. Reprinted as Commentaries on the History and Cure of Diseases. Birmingham, AL: The Classics of Medicine Library, Division of Gryphon Editions, Ltd., 1982: 395–397.

Who named it?

It was not until 1837 that Johann Schönlein recognized a similar constellation of symptoms, and named the arthritic component associated with purpura "peliosis rheumatica" or "purpura rubra."

Schönlein JL. Allgemeine und Specielle Pathologie undTherapie, Vol. 2, 3rd edn. Wurzburg: Herisau, 1837: 48.

Who named it?

Schönlein, in addition to the other symptoms, also described renal involvement, citing “frequent precipitates in the urine” of patients.

Schönlein JL. Allgemeine und Specielle Pathologie undTherapie,

Vol. 2, 3rd edn. Wurzburg: Herisau, 1837: 48.

Who named it?

It was Eduard Heinrich Henoch, a pupil of Schönlein, who added abdominal colic, bloody diarrhea, and hemorrhagic nephritis to the syndrome's components.

Henoch EH. Über eine eigenthumliche Form von Purpura. Berl Klin Wochenschr 1874; 11: 641–643.

Epidemiology

- It is most common in children
- Often associated with an inciting infection, such as group A streptococcus or other exposure
- There is a male predominance
- Occurs most frequently in spring and summer

Kraft DM, et al. Henoch-Schönlein purpura: a review. Am Fam Physican. 1998;58(2):405-408,411.

Possible Etiologies

- Upper Respiratory Tract Infections
- Streptococcal infections
- Other infections
- Vaccinations
- Insect Bites

Kliegman RM, et al.
Nelson Textbook of Pediatrics. 19th ed.
Philadelphia, Pa.: Saunders Elsevier; 2011.

Precipitating Antigens

- INFECTIONS
  - URI
  - Measles
  - Rubella
  - Parvovirus B19
  - Mycoplasma
  - Coxsackie virus
  - Toxocara
  - Amebiasis
  - Salmonella
  - C.difficile
  - H.pylori
  - Adenovirus
  - Legionella
  - Tuberculosis
  - Mumps
  - Streptococcus
  - Morganella morganii

Precipitating Antigens

Drugs

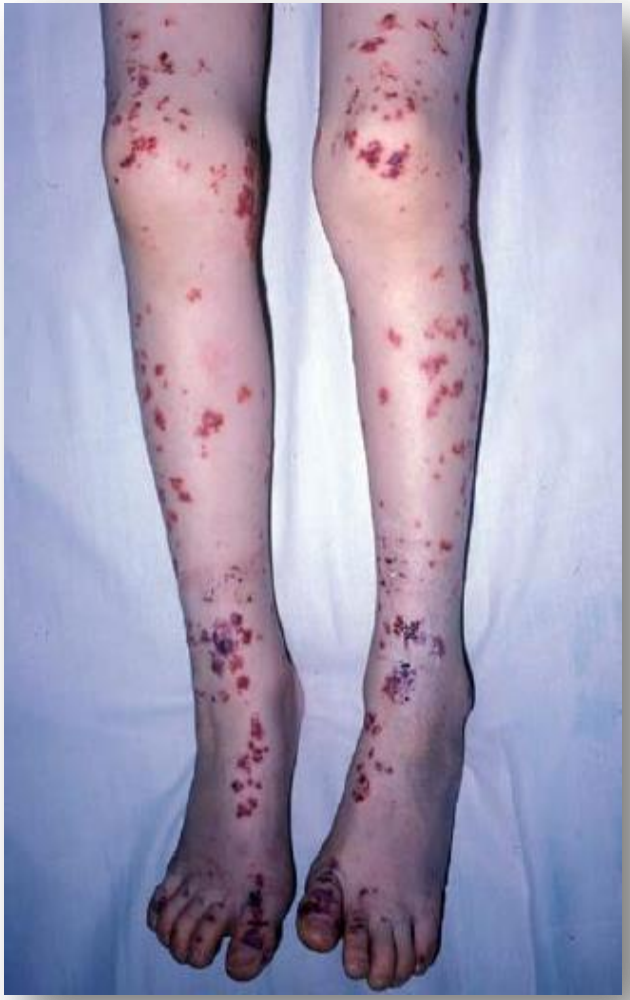
- Vancomycin
- Streptokinase
- Ranitidine
- Cefuroxime
- Diclofenac
- Enalapril
- Captopril

Kliegman RM, et al.
Nelson Textbook of Pediatrics. 19th ed.
Philadelphia, Pa.: Saunders Elsevier; 2011

Clinical Features

Tetrad of symptoms

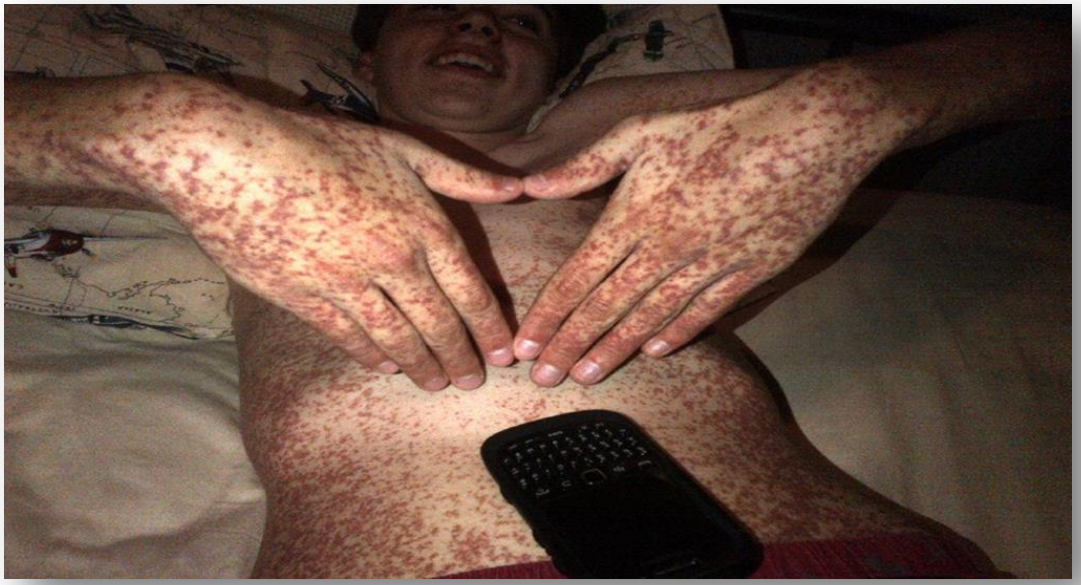
- Palpable purpura
- Hematuria
- Arthritis/arthralgias
  - more common in adults
  - most common in knees and ankles.
  - Generally self-limiting
- Abdominal pain
  - less frequently in adults than in children.



Clinical Features

PALPABLE PURPURA: most commonly seen on lower extremities and buttocks, however can also been seen on the trunk and arms.

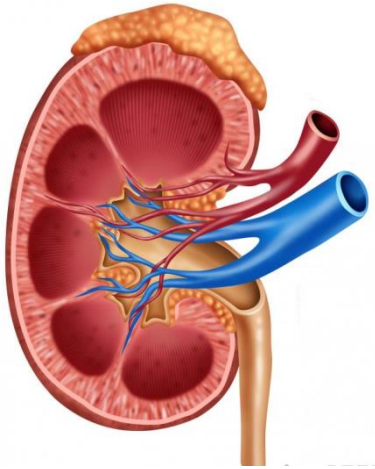
- Lesions begin as erythematous macules and progress to purpuric, non-blanching, nonpruritic lesions that may become confluent



Clinical Features

RENAL INVOLVEMENT:

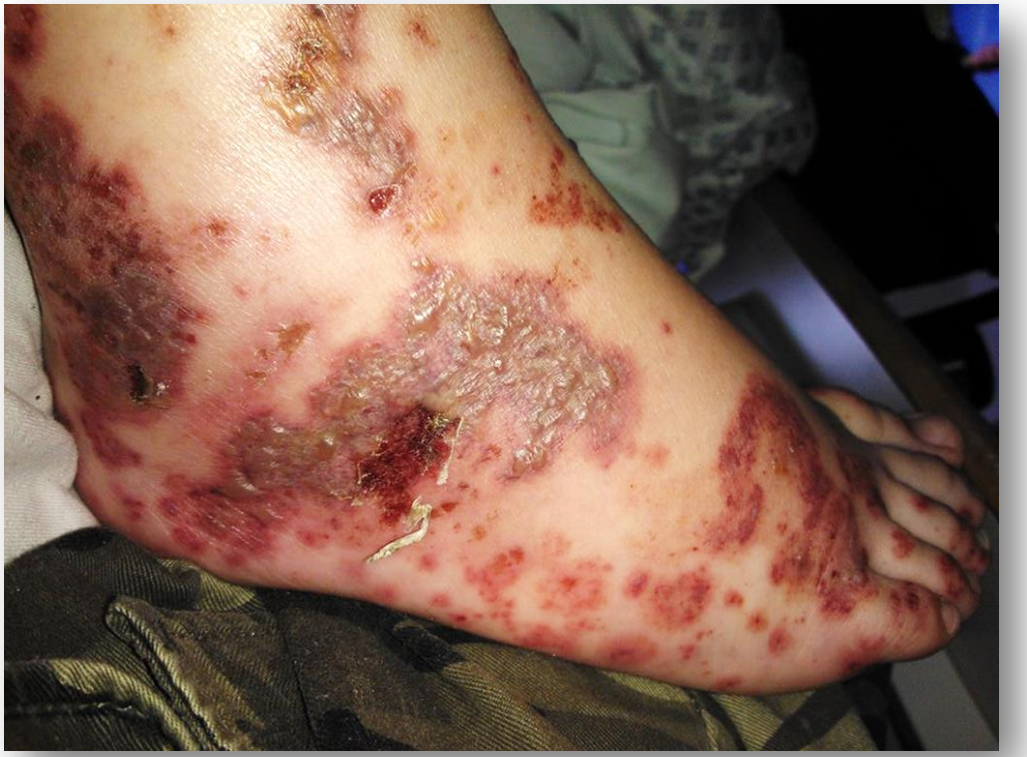
- in up to 50% of patients
- Usually more rapidly progressive in adults. Rare in children
- May present with hematuria
- Can have mild glomerulonephritis leading to microscopic hematuria and can lead to a rapidly progressive glomerulonephritis with RBC casts
- Usually resolve spontaneously.



Schienfeld NS, Jones EL. Pediatric Henoch-Schönlein purpura Medscape Reference. http://www.emedicine.medscape.com/article/

Clinical Features

Joint symptoms are present in the majority of patients, the knees and ankles being most commonly involved.



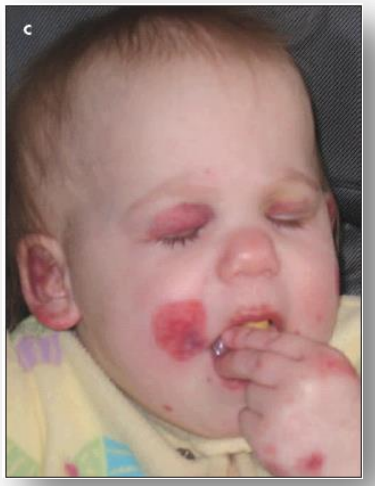
Clinical Features

- Abdominal pain secondary to vasculitis of the intestinal tract is often associated with gastrointestinal bleeding.

Davin JC. Henoch-Schönlein purpura nephritis: pathophysiology, treatment, and future strategy. Clin J Am Soc Nephrol. 2011 Mar;6(3):679-89.

Clinical Findings

- Acute hemorrhagic edema of childhood (AHEC) is an acute leukocytoclastic vasculitis which affects children under the age of 2 years.
- AHEC and HSP share features such as the prodrome, seasonal predisposition, cutaneous involvement, and histologic findings on skin biopsy. In AHEC, facial edema may be the initial sign



Shah D, Goraya JS, Poddar B, et al. Acute infantile hemorrhagic edema and Henoch–Schönlein purpura overlap in a child. Pediatr Dermatol 2002; 19: 92–93

Laboratory Findings

Generally a clinical diagnosis
There isn't a single test to confirm HSP

- May have mild leukocytosis
- Normal platelet count
- Normal serum complement levels
- Elevated IgA in 50%

Davin JC. Henoch-Schönlein purpura nephritis: pathophysiology, treatment, and future strategy. Clin J Am Soc Nephrol. 2011 Mar;6(3):679-89.

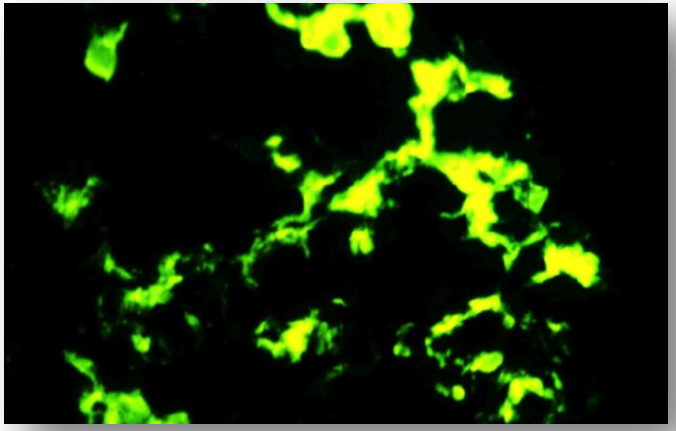
Laboratory Findings

- HSP is characterized by leukocytoclastic vasculitis with serum immunoglobulin A (IgA) deposition in vessel walls.
- This deposition in the small vessels through the body leads to the hallmark symptoms of HSP.
- IgA depositions accumulating in the vessels of the intestinal mesentery result in HSP's gastrointestinal manifestations.

Saulsbury FT. Epidemiology of Henoch-Schönlein purpura . *Cleve Clin J Med.* 2002;69(suppl 2):SII87-SII89

Laboratory Findings

- Skin Biopsy: can be helpful and used to confirm IgA and C3 deposits and leukocytoclastic vasculitis.
- Renal Biopsy: not usually needed for diagnosis. Will show mesangial IgA deposits and segmental glomerulonephritis.



Saulsbury FT. Henoch-Schönlein purpura in children: report of 100 patients and review of the literature. Medicine 1999; 78: 395-409

Laboratory Findings

- A decrease in GFR is common with a nephritic presentation
- The renal lesions are considered by most experts to be identical to those found in IgA nephropathy
- Most patients with microscopic hematuria and minimal proteinuria recover fully over several weeks

Coppo R, Peruzzi L, Amore A et al (2007) IgACE: a placebo controlled, Randomized trial of angiotensin-converting enzyme inhibitors in children and young people with IgA nephropathy and moderate proteinuria. J Am Soc Nephrol 18(6):1880-1888.

Complications

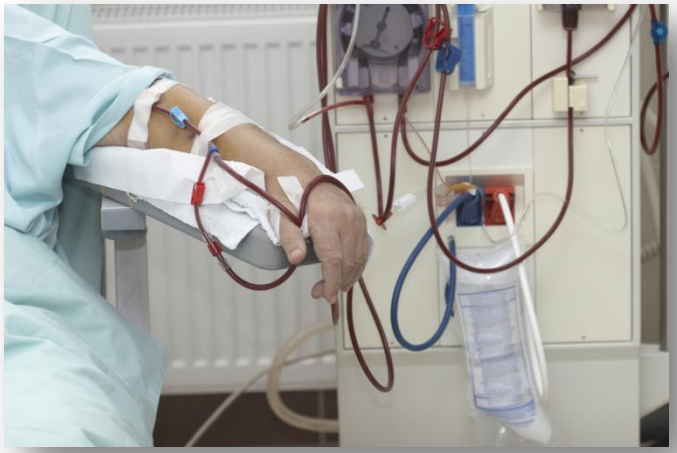
Renal Involvement- Children

- Only 1-5% progress to ESRD.
- 10-50% of children get microscopic hematuria, mild GN, and proteinuria that resolves spontaneously.
- Up to 33% recurrence in children, but symptoms are milder and shorter duration.

McCarthy HJ et al. Clinical practice: diagnosis and management of Henoch-Schönlein purpura. Eur J Pediatr. 2010 Jun;169(6): 643-50.

Complications

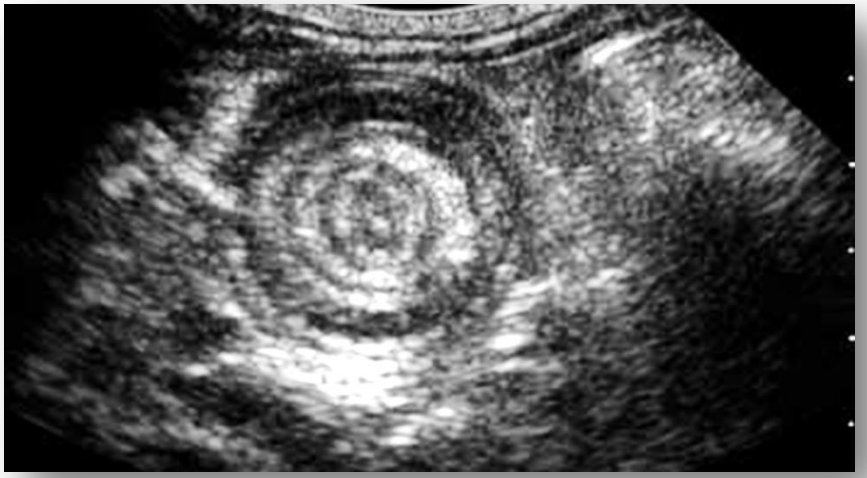
- Progressive CKD and possibly ESRD are more likely to develop in those with the nephrotic syndrome and the presence of both nephritic and nephrotic syndrome poses the worst renal prognosis.
- Occasionally the damage is severe enough that dialysis or a kidney transplant may be needed.



Xia Y et al.
Clinical outcomes in children with Henoch-Schönlein purpura nephritis grade IIIa or IIIb. Pediatr Nephrol. 2011 Jul;26(7):1083-8

Complications

Bowel obstruction. In rare cases, Henoch-Schönlein purpura can cause intussusception — a condition in which a section of the bowel folds into itself like a telescope, which prevents matter from moving through the bowel.



Ronkainen J: The adult kidney 24 years after childhood HSP: retrospective cohort. Lancet 360: 666-670, 2002.

Supportive Treatment of HSP

- Most patients may be treated on an outpatient basis
- Advise patients to rest until symptoms wear off
- Prognosis is generally good, especially if no renal involvement
- STRICT Follow-up should be advised

Interventions for preventing and treating kidney disease in Henoch–Schönlein Purpura (HSP) (Review)

THE COCHRANE COLLABORATION®

Treatment of HSP

- There are few data from randomized studies examining interventions used to prevent or treat serious kidney disease in HSP except for short-term prednisone to prevent kidney disease.
- There was no evidence of benefit of prednisone compared with placebo or no specific therapy in preventing serious kidney disease in HSP.

THE COCHRANE COLLABORATION®

Treatment of HSP

NSAIDS:

- increased risk of GI bleed

Steroids:

- Prednisone 1 mg/kg
- Helps with arthralgias and abdominal sxs
- not proven to benefit skin or renal disease
- Does not shorten duration of active disease
- Does not prevent recurrence

Severe renal disease: 'limited evidence'

- cyclophosphamide,
- Plasmapharesis
- IVIG
- cyclosporin

Criteria for Hospitalization

1. Inability to maintain adequate hydration orally
2. Severe anemia requiring transfusion
3. Severe abdominal pain
4. Significant GI bleeding
5. Changes in mental status
6. Severe joint involvement limiting ability to move
7. Renal insufficiency, hypertension and nephrotic syndrome

Treatment of HSP

Emerging Therapies

- **Dapsone**

  A bacteriostatic drug with anti-inflammatory properties that may hasten the resolution of the palpable rash. Further studies are needed.

- **Methotrexate**

  Immunosuppressive agent. Insufficient data in HSP.

- **ACE inhibitors**

  May help reduce proteinuria and protect the kidney in HSP; however, there are insufficient data and more testing is needed.

Zaffanello M, Brugnara M, Franchini M. Therapy for children with Henoch-Schönlein purpura nephritis: a systematic review. S ScientificWorldJournal. 2007;7:20-30

Treatment of HSP

- **Mycophenolate**

  Immunosuppressive agent. Insufficient data in HSP, needs further testing.

- **Urokinase**

  Anticoagulant. Insufficient data in HSP, needs further testing.

- **Intravenous immunoglobulin (IVIG)**

  May help reduce proteinuria; however, IVIG may also be nephrotoxic. No convincing studies to date.

- **Factor XIII administration**

  May help treat severe GI tract bleeding complications; however, there are no convincing studies to date.

Treatment of HSP

- **Tonsillectomy**

  Tonsils may be a source of abnormal IgA that forms immune complexes. Requires further study.

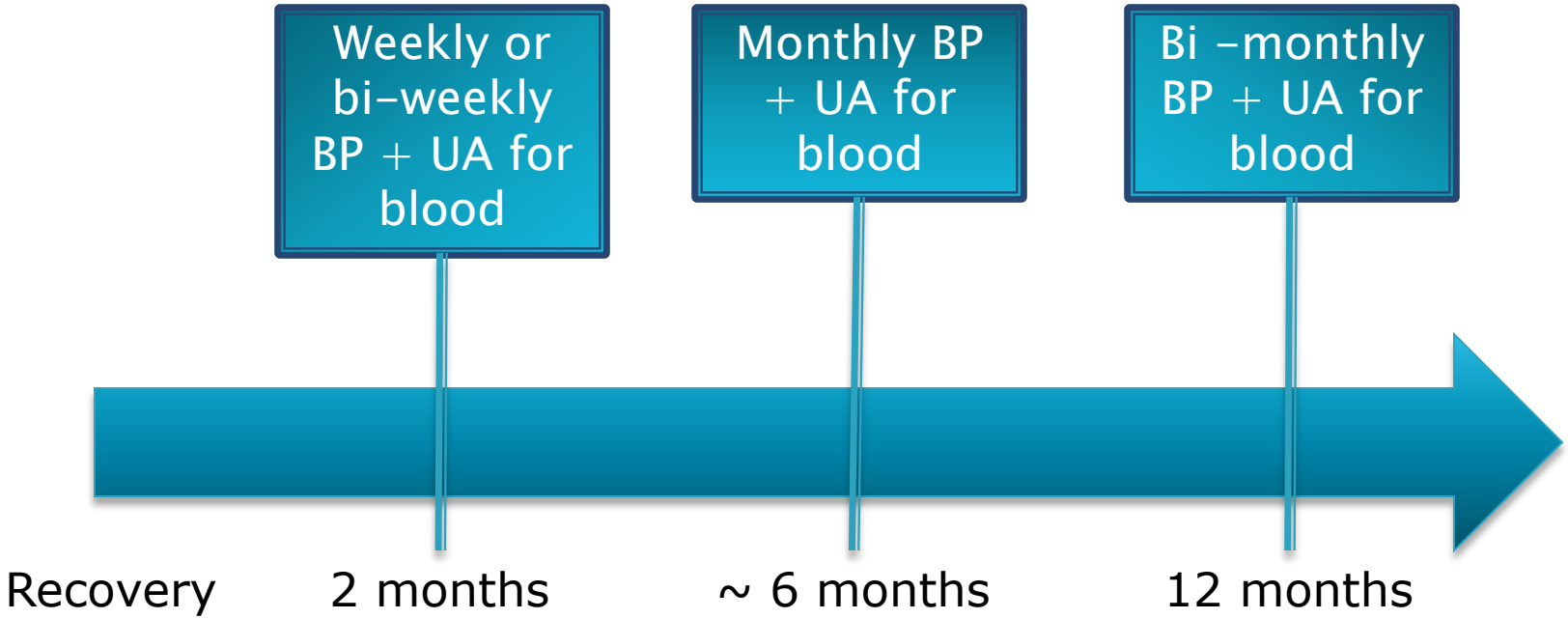
- **Vitamin E**

  There are no convincing studies to date.

- **Fish oil**

  May have anti-inflammatory properties; however, No convincing studies to date.

Follow-up



Obtain SERUM CREATININE anytime if (+) abnormalities

Sources

- Ronkainen J: The adult kidney 24 years after childhood HSP: retrospective cohort. Lancet 360: 666-670, 2002
- Pankhurst T: Malignancy is increased in ANCA assoc vasculitis, Rheumatology 43:1532, 2004
- Coppo R: Long term Prognosis of HSP nephritis in adults and children. Nephrol Dial Transplant 12: 2277-2283, 1997
- Evangeline, P: HSP in adults, Outcome and Prognosis factors, J Am Soc Nephrology 13:1271: 2002