HENOCH SCHONLEIN PURPURA CLINICAL PATHWAY
PHOENIX CHILDREN'S HOSPITAL

PATHWAY FLOW DIAGRAM/ALGORITHM
See Figures 1, 2, and 3

STANDARD DISCLAIMER
This guideline is not intended to replace clinical judgment. It is meant to assist licensed independent practitioners and other health care providers in clinical decision-making by describing a range of generally acceptable approaches to the diagnosis and management of a particular condition. A particular patient's circumstances should always be taken into account when a practitioner is deciding on a course of management.

SCOPE
Definition

- Pediatric patients (age <16 years) presenting with physician diagnosed Henoch Schonlein Purpura (HSP) to Phoenix Children's Hospital (PCH) or Phoenix Children's Care Network (PCCN) facility.

Inclusion Criteria

1. Pediatric patients age <16 years
2. Palpable purpura or petechiae predominantly of the lower extremities and the buttocks (100% of patients)
3. Supportive criteria NOT NECESSARY for inclusion:
    a. Diffuse, acute colicky abdominal pain (67% of patients)
    b. Arthritis and/or arthralgias (50-80% of patients)
    c. Hypertension (3% of patients)
    d. Scrotal pain and swelling in males (2-38% of patients)

Exclusion Criteria

- Age > 16 years
- Thrombocytopenia and its causes
    a. Leukemia and malignancy
    b. Idiopathic Thrombocytopenic Purpura
    c. Hemolytic Uremic Syndrome or Thrombotic Thrombocytopenic Purpura
- Disseminated intravascular coagulation
- Evidence of sepsis (especially meningococcal sepsis)
- Other forms of vasculitis:
    a. Cutaneous small vessel vasculitis
    b. ANCA-associated vasculitis (including granulomatosis with polyangiitis, microscopic polyangiitis)
    c. Systemic lupus erythematosus
    d. Polyarteritis nodosa
- Inflammatory bowel disease

KEY CLINICAL RECOMMENDATIONS

1. Pediatric patients presenting in the outpatient setting with suspicion for HSP (rash, joint pain, joint swelling, and abdominal pain) should be evaluated with an in-office urine dipstick and be sent for the following labs: CBC, ESR/CRP, CMP, PT/PTT, UA and random urine protein and creatinine levels.
2. Patients who are ill-appearing or having difficulty ambulating should be referred directly to the Emergency Department for further evaluation.
3. Low grade fever is a common presenting sign in patients with HSP; however, the presence of fever should alert the provider to consider alternative diagnoses such as sepsis and Rocky Mountain Spotted Fever.
4. Weekly urinalysis, urine protein/creatinine ratios, and blood pressure should be recorded for the first 4 weeks after onset of the disease- if these are abnormal, a repeat blood pressure and a first morning void sample with a UA, urine protein and urine creatinine testing should be performed. A referral to pediatric nephrology should be considered if there is persistent hypertension and/or abnormal testing on the first AM void sample including macroscopic hematuria, microscopic hematuria (5 or more RBCs per high powered field) or proteinuria (>0.2 mg/mg creatinine or 200 mg/gm creatinine).
5. Monthly urinalysis, urine protein/creatinine ratios, and blood pressure should be recorded for the first 6 months after onset of the disease- if these are abnormal, a repeat blood pressure and a first morning void sample with a UA, urine protein and urine creatinine testing should be performed. A referral to pediatric nephrology should be considered if there is persistent hypertension and/or abnormal testing on the first AM void sample including macroscopic hematuria, microscopic hematuria (5 or more RBCs per high powered field) or proteinuria (>0.2 mg/mg creatinine or 200 mg/gm creatinine).
6. Pediatric patients presenting to the PCH Emergency Department with suspicion for HSP (rash, joint pain, joint swelling, and abdominal pain) should be evaluated with the following: CBC, ESR/CRP, CMP, PT/PTT, UA and random urine protein and creatinine levels, and an abdominal US if significant abdominal pain or other concerning symptoms of intussusception.
7. Patients with signs/symptoms of significant HSP-related GI disease (abnormal US, presence of GI hemorrhage, or severe abdominal pain- i.e. pain that limits normal activities AND is associated with intolerance for adequate hydration and/or nutrition) should have the following consults considered: Pediatric surgery, Pediatric gastroenterology, and Pediatric rheumatology.
8. Patients with signs/symptoms of significant HSP-related renal disease (hypertension, urine protein:creatinine ratio >0.2 mg/mg creatinine or 200 mg/gm creatinine), macroscopic hematuria, microscopic hematuria (5 or more RBCs per high powered field), elevated serum creatinine level, hypoalbuminemia, peripheral edema) should have the following consult considered: Pediatric nephrology.
9. Patients with signs/symptoms of significant HSP-related joint or skin disease (severe joint pain, inability to ambulate, or blistering/ulcerating skin rash) should have the following consult considered: Pediatric rheumatology.
    a. Follow up with specialists arranged as necessary
10. Supportive therapy should be considered for all patients with HSP:

a. NPO/IV rehydration as necessary (consider consultation with pediatric nephrology if patient edematous and/or evidence of renal disease)
b. Encourage PO fluids
c. NSAIDs or other analgesics

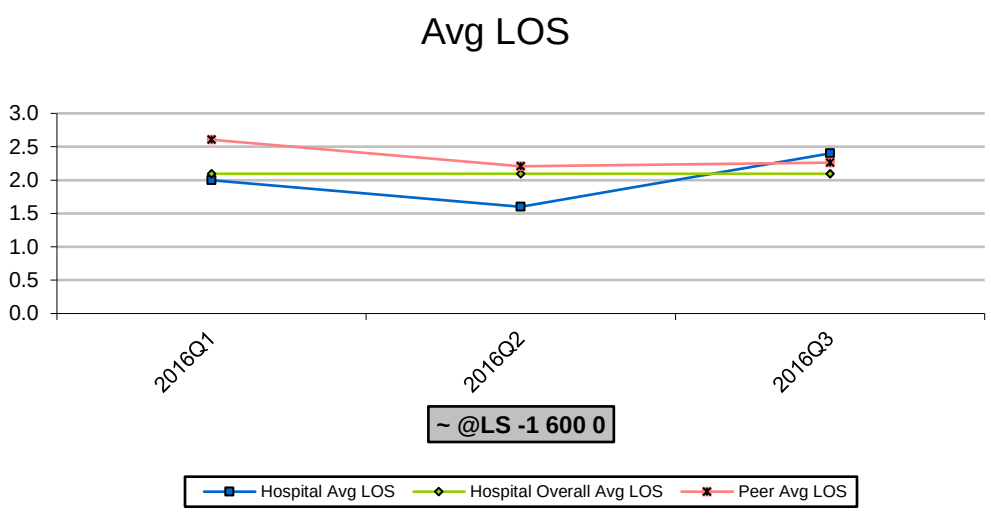
11. Oral or IV corticosteroid agents should not be routinely used in patients with HSP and consultation with Pediatric Rheumatology should be considered in patients who may require corticosteroids.

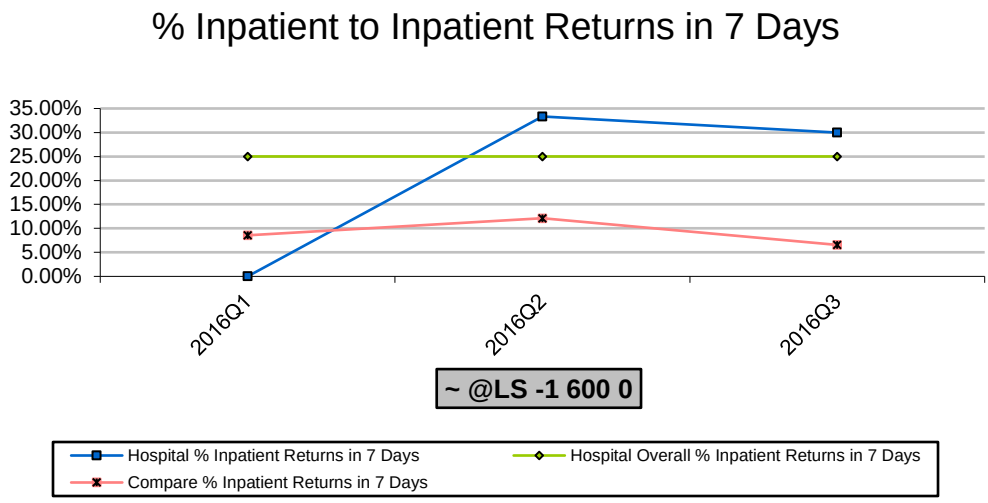
PATHWAY GOALS

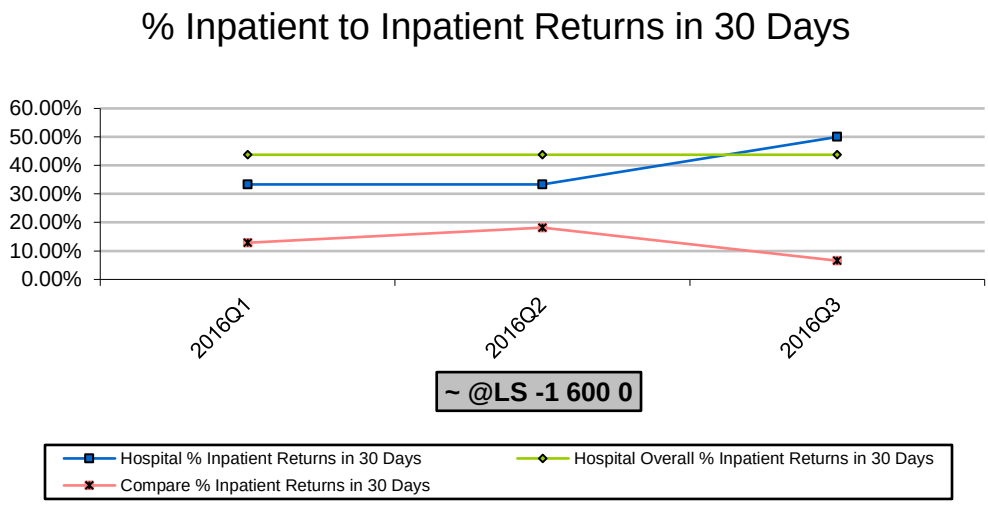
1. Standardize the evaluation and treatment of patients with HSP.
2. Decrease the number of referrals for patients with HSP to the Emergency Department who are identified as low risk for complications.
3. Decrease the number of patients with HSP admitted to PCH from the Emergency Department who are identified as low risk for complications.
4. Reduce the average length of stay of patients with HSP admitted to PCH.
5. Reduce the rate of readmission of patients with HSP at PCH both at 7 and 30 days.

BASELINE DATA

- Baseline data comparing PCH to PHIS from Q1-3 of 2016
- # Admits for HSP: PCH 21, PHIS 10
- Average LOS: PCH 2.01, PHIS 2.39
- 7 day readmission rate: PCH 23.81%, PHIS 9.79%
- 30 day readmission rate: PCH 42.86%, PHIS 18.18%
- Steroid use: PCH 48%, PHIS 55%







OUTCOME/PROCESS/BALANCING MEASURES

1. Outcome Measures
    a. Outpatient
        i. Ability to obtain outpatient follow-up in recommended time frame
    b. Emergency Department
        i. Hospitalization rate
        ii. Steroid use
    c. Hospital
        i. Average length of stay
        ii. Steroid use
2. Process Measures
    a. Outpatient
        i. Renal monitoring schedule
    b. Emergency Department
        i. Use of clinical pathway/order set

ii. Ability to make a PCP appointment within 1 week of discharge

c. Hospital
   i. Use of clinical pathway/order set
   ii. Ability to make a PCP appointment within 1 week of discharge

3. Balancing Measures
   a. Outpatient
   b. Emergency Department
      i. Number of patients with missed renal involvement
      ii. Number of patients with missed intussusception
      iii. 7 day return to emergency department rate
      iv. 30 day return to emergency department rate
   c. Hospital
      i. Number of patients with missed renal involvement
      ii. Number of patients with missed intussusception
      iii. 7 day readmission rate
      iv. 30 day readmission rate

ADMISSION CRITERIA

1. Patient with confirmed HSP AND ANY OF THE FOLLOWING:
   a. Hypertension
   b. Significant edema
   c. Abnormal serum creatinine level
   d. Severe joint pain/inability to ambulate
   e. Severe abdominal pain
   f. GI hemorrhage
   g. Abnormal abdominal imaging (e.g. intussusception on US)
   h. Other concerning findings (e.g. orchitis, neurologic symptoms)

DSICHARGE CRITERIA

1. Tolerating PO intake for >24 hrs prior to discharge
2. No requirement of IV analgesics for >24 hrs prior to discharge
3. No hypertension or well controlled hypertension
4. Ambulating safely
5. Follow up with specialists arranged as necessary

ORDER SET

See “HSP Clinical Pathway Order Set” attachment

PATIENT AND FAMILY EDUCATION / DISCHARGE PLANNING

1. Handouts:
   a. English: https://www.printo.it/pediatric-rheumatology/GB/info/pdf/8/Henoch-Schoenlein-Purpura
   b. Spanish: https://www.printo.it/pediatric-rheumatology/ES_ES/info/pdf/8/P%C3%BArpura-de-Sch%C3%B6nlein-Henoch

EVIDENCE BASED SUPPORTING MATERIAL

Henoch Schonlein Purpura (HSP) is the most common childhood vasculitis with an estimated incidence of 20 per 100,000 children per year.[1] HSP results in visits to primary care physicians, urgent cares/emergency departments and ultimately inpatient admissions. The evaluation, treatment and management of HSP varies widely across the continuum of pediatric care. This clinical pathway aims to standardize the evaluation and treatment of HSP within Phoenix Children's Hospital and Phoenix Children's Care Network.

The classification criteria for HSP were updated in 2010 and while they were developed to enroll patients in clinical research studies, they can be used to aide clinicians in the diagnosis of patients with HSP.[2] HSP features a wide variety of clinical manifestations including skin, joint, abdominal, renal and other rare complications. While the vast majority of patients recover without any significant long-term complications, serious and potentially life-threatening renal disease occurs in less than 10% of patients, and intussusception occurs in less than 5% of patients.[3]

There are no current evidenced-based guidelines for the evaluation and management of children with HSP. There are some clinical pathways available on the internet from other institutions with some evidence to support the specific recommendations.[4] The Cochrane Library did perform a systematic review of treatment for preventing and treating kidney disease in HSP and found that prednisone did not prevent persistent kidney disease from developing.[5] Corticosteroids may have a role in the hospitalized patient in relation to the gastrointestinal manifestations of the disease, as well as in severe joint or skin manifestations.[3,6]

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6. Weiss PF, Klink AJ, Localio R, Hall M, Hexem K, Burnham JM, Keren R, Feudtner C. Corticosteroids may improve clinical outcomes during hospitalization for Henoch-Schönlein purpura. Pediatrics. 2010 Oct;126(4):674-81.

DEVELOPMENT AND APPROVAL, AND IMPLEMENTATION PROCESS

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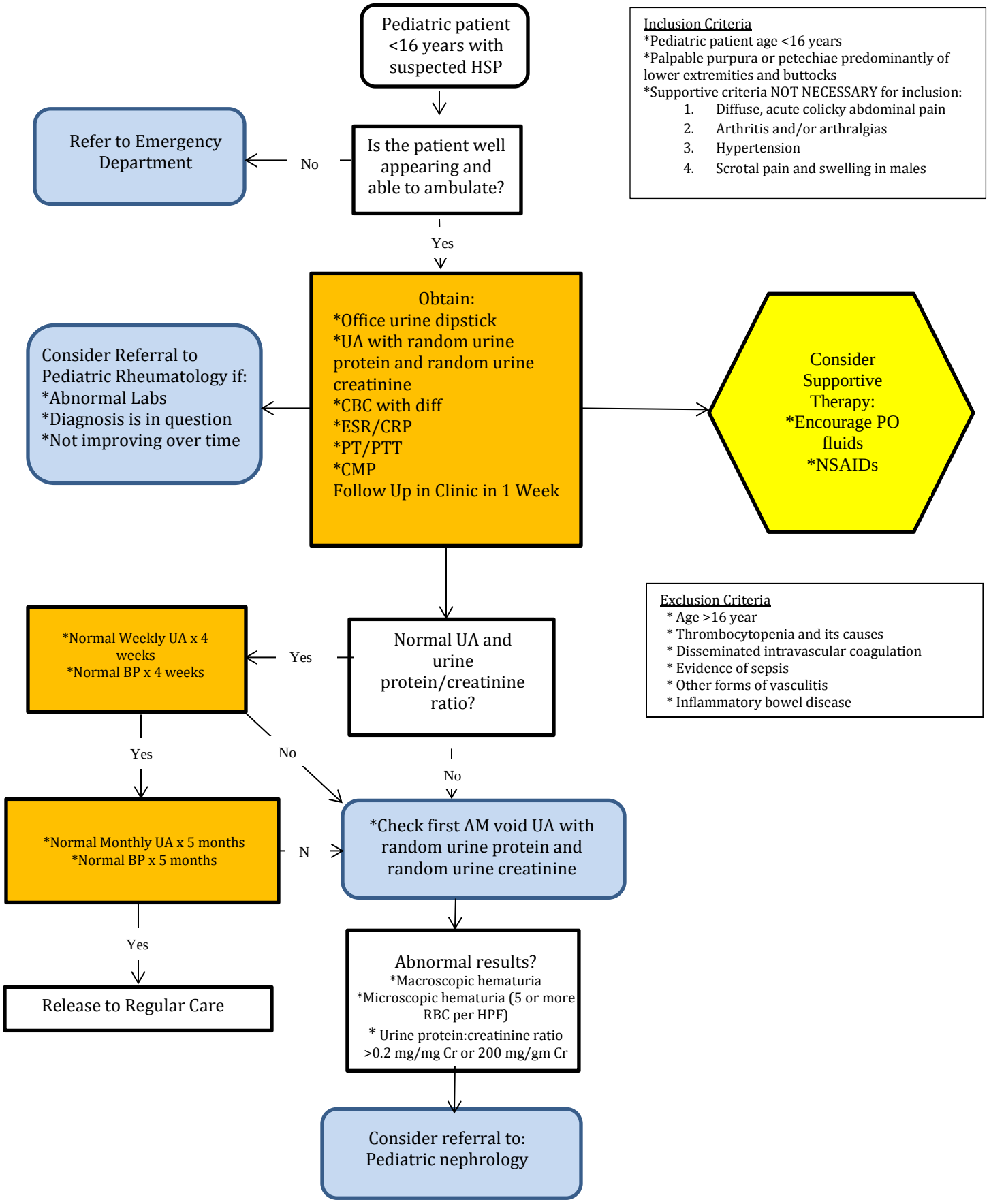
- Additional Stakeholders: Sections of General Pediatrics (Kristen Samaddar, MD) Nephrology (Karen Papez, MD), Gastroenterology (Mark McOmber, MD) and Surgery (Justin Lee, MD)
- Approved by Clinical Standards Committee: March 2018
- Approved by Clinical Effectiveness Committee: April 2018

Figure 1 – Out-Patient HSP Algorithm



Inclusion Criteria
*Pediatric patient age <16 years
*Palpable purpura or petechiae predominantly of lower extremities and buttocks
*Supportive criteria NOT NECESSARY for inclusion:
1. Diffuse, acute colicky abdominal pain
2. Arthritis and/or arthralgias
3. Hypertension
4. Scrotal pain and swelling in males

Exclusion Criteria
* Age >16 year
* Thrombocytopenia and its causes
* Disseminated intravascular coagulation
* Evidence of sepsis
* Other forms of vasculitis
* Inflammatory bowel disease

Figure 2 – Emergency Department HSP Algorithm

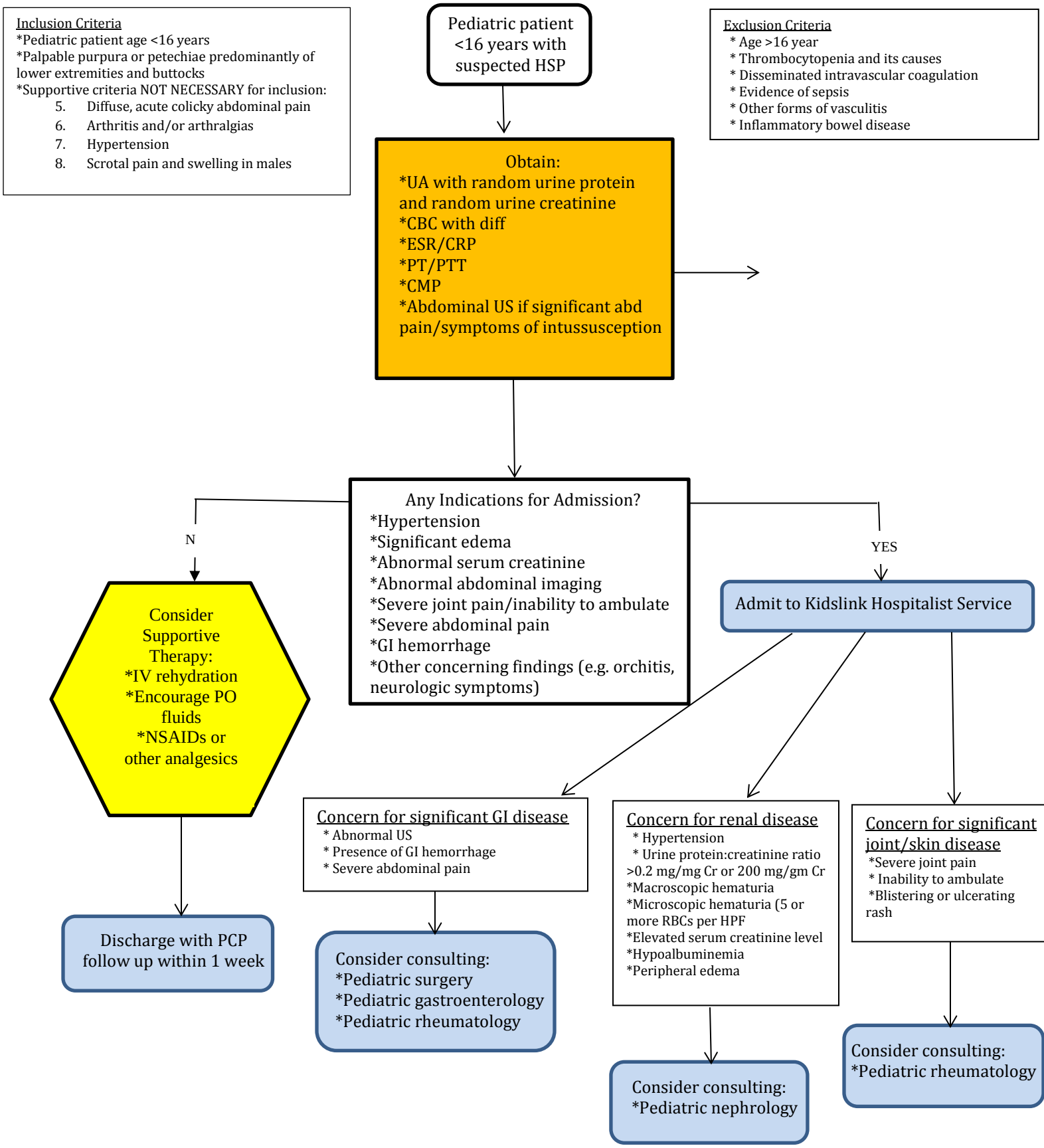


Figure 3 – In-Patient HSP Algorithm