CLINICAL GUIDELINE

IgA Vasculitis Clinical Guideline
Emergency Department/Outpatient Evaluation and Management

Children's MINNESOTA

Aim: To standardize the evaluation and management of patients with IgA vasculitis

Exclusion guidelines:
- Toxic-appearing or septic
- Age <2 years old
- History of chronic kidney disease
- History of inflammatory bowel disease
- History of prior IgA Vasculitis
- Thrombocytopenic patients

Patient with concern for IgA vasculitis:
Purpuric/petechial rash (required) with abdominal pain and/or joint pain
(see note 1 for an overview of clinical presentation and diagnosis of IgA vasculitis)

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Obtain the following initial workup: Urinalysis, blood pressure (see note 3), CBC with differential, and renal panel (BMP, Phos)

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Consider additional work-up (if concerns or diagnosis is unclear):
- Urine protein:creatinine ratio (if urine dipstick has ≥2+ protein)
- CRP, ESR, INR/PTT, Lyme panel, lactate, lipase
- Stool pathogen panel and/or c. diff testing (if hematochezia and diagnosis unclear)
- Limited abdominal ultrasound to screen for intussusception (if severe abd pain)
- Testicular ultrasound (if testicular pain or scrotal swelling)
- Consider rheumatology consult (Box 2) if atypical presentation (see note 1)

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Does the patient have signs of any clinical complications of IgA vasculitis? (see note 2)

Yes → **If outpatient, send to ED. If in the ED, Admit to Hospital**
See p. 2 for Inpatient Management

No ↓

Does the patient meet admission criteria (Box 1)?
Call nephrology ASAP if renal dysfunction is so severe that patient may need transfer to a hemodialysis center

Yes → **Admit to Hospital**
See p. 2 for Inpatient Management

No → **Discharge Patient**
- See note 4 for Follow-up Recommendations
- See note 5-6 for pain control
- See note 8 for prognostic information and anticipatory guidance

Box 1- Admission Criteria
- Recommended by subspecialist
- Acute kidney injury (i.e. elevated creatinine)
- Hypertension requiring acute treatment
- Nephrotic syndrome requiring diuretics
- Clinically unwell or dehydrated
- Severe pain requiring IV analgesics
- Gastrointestinal bleed
- Neurological symptoms
- Concern for alternate etiologies requiring additional workup

Box 2- Rheumatology Consult Process
- Call Children's Physician Access (**612-343-2121**) and request a call to UMN pediatric rheumatology for a consult
- Direct UMN provider referral line if needed: 1-888-543-7866

CLINICAL GUIDELINE

IgA Vasculitis
Inpatient Evaluation and Management

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Patient is Admitted with Suspected IgA Vasculitis

Evaluate for Clinical Complications (see note 2)

1. Pain control: See note 5
   - NSAIDs and acetaminophen as first line.
   - Consider systemic steroids for debilitating pain (notes 5 and 6). If initiating steroids, consult Endocrinology to teach stress coverage pending outpatient follow-up to assess for adrenal insufficiency.
2. Consult nephrology if indicated (see note 7)
3. Consult gastroenterology if unremitting abdominal pain, GI bleed, or any GI complications (see note 2)
4. Closely monitor blood pressures (note 3), fluid status including strict I/Os, daily weights.
5. Monitor urinalysis and renal panel if initial were abnormal or if new concerns for renal function develop (e.g. hypertension, decreased urination)
   - If urinalysis shows ≥2+ proteinuria, obtain a spot urine protein:creatinine ratio
6. Consider additional work-up if not obtained prior to admission or new concerns arise- see p.1 suggestions for further potential work-up
   - If presentation unclear or clinical course not as expected, call rheumatology at the UMN (see p.1, Box 2)
     - May consider: ASO titers and anti-DNASe B, ANA dsDNA, ANCA, C3, C4, immunoglobulins
     - If patient is of Mediterranean descent or has a history of periodic fevers, consider discussing an evaluation for Familial Mediterranean Fever, which can be associated with IgA Vasculitis (testing includes serum amyloid level and MEFV gene mutation testing)

Does the patient meet discharge criteria?

- Cleared by consultants (if they were involved in patient's care)
- If hypertensive, not requiring IV anti-hypertensive medication for >24 hours
- Creatinine normal or stable
- Clinically improving
- Pain well-controlled on oral analgesia
- No complications of IgA vasculitis that require ongoing inpatient management

Yes → **Discharge Patient**

See note 4 for Follow-up Recommendations

No → **Consider rheumatology consultation (see p.1 Box 2)**

(may need to transfer to a facility with inpatient rheumatology)

Aim: To standardize the evaluation and management of patients with IgA vasculitis

Note 1: IgA vasculitis overview and clinical presentation

- IgA vasculitis, formerly known as Henoch-Schönlein purpura (HSP), is the most common vasculitis of childhood. Typically affects children 2-8 years old.
- Recent upper respiratory tract infection is present in 50% of cases, commonly viral or Group A streptococcus infections
- Most cases are self-limited and only require symptomatic management
- IgA vasculitis is a *clinical diagnosis* based on a patient having a classic **rash** (petechial or purpuric, symmetrical, gravity/pressure dependent areas, eventually seen in 100% of patients) AND at least one of the following:
    - Arthritis/arthralgias (50-75%): usually affects large joints of lower limbs, rarely effusion or warmth. May simply present as "decreased ambulation" in a younger child
    - Abdominal pain (50%)
    - Nephritis (25-50%, may be delayed up to 6 months after rash): may include proteinuria, hematuria, nephrotic syndrome, acute nephritic syndrome, hypertension, renal impairment
    - Histopathological confirmation (typically skin biopsy showing leucocytoclastic vasculitis with predominant IgA deposit or proliferative glomerulonephritis with predominant IgA deposit)
- Clinical manifestations can take days to weeks to fully develop, may vary in order of appearance
- Children >12 years of age have a significantly increased risk of renal involvement, developing corticosteroid dependence, and developing refractory disease
- Labs may show leukocytosis and eosinophilia, however platelets and coagulation studies (if obtained) should be normal in IgA vasculitis and, if abnormal, alternative diagnoses should be sought.
- Differential diagnosis for a patient presenting with a petechial/purpuric rash and joint pains or abdominal pain includes but is not limited to: other vasculitis (e.g. granulomatosis with polyangiitis, polyarteritis nodosa), acute hemorrhagic edema of infancy (patients <3 years old), sepsis, leukemia, causes of thrombocytopenia such as immune thrombocytopenic purpura and hemolytic uremic syndrome.

Note 2: Complications of IgA vasculitis that would warrant subspecialist involvement

- GI: intussusception, GI hemorrhage, bowel ischemia, necrosis or perforation, protein-losing enteropathy, pancreatitis
- Renal: nephritic or nephrotic syndrome, hypertension, renal failure
- Derm: painful non-pitting subcutaneous edema
- Pulm: diffuse alveolar hemorrhage, interstitial disease
- Neuro: CNS vasculitis (may include brain hemorrhages, infarcts and edema), posterior reversible encephalopathy syndrome and hypertensive encephalopathy, seizures
- Urology: orchitis, testicular necrosis

Note 3: Evaluating for hypertension

- Accurately measuring blood pressure: patient should be seated, in a quiet room, back supported, feet flat, measured on the right arm, middle of the cuff at heart level. If automatic BP reading meets hypertension criteria, make sure cuff is the right size and confirm with a manual reading.
- Definition of hypertension:
    - Age <13 years: systolic or diastolic BP >95th percentile for age, sex and height according to the **table on p.4**
    - Age ≥ 13 years: ≥130/80
- Acute treatment of hypertension should be determined in consultation with nephrology (and may be impacted by factors such as patient's pain control and use of systemic steroids)

Aim: To standardize the evaluation and management of patients with IgA vasculitis

Note 3 Continued- Tables Defining Hypertension *(adapted from the Pediatrics 2017 "Clinical Practice Guideline for Screening and Management of High Blood Pressure in Children and Adolescents")*

		SBP (mm Hg)							DBP (mm Hg)						
		Height Percentile or Measured Height							Height Percentile or Measured Height						
Age (y)	BP Percentile	5%	10%	25%	50%	75%	90%	95%	5%	10%	25%	50%	75%	90%	95%
	Height (in)	30.4	30.8	31.6	32.4	33.3	34.1	34.6	30.4	30.8	31.6	32.4	33.3	34.1	34.6
1	95th	102	102	103	103	104	105	105	54	54	55	55	56	57	57
	Height (in)	33.9	34.4	35.3	36.3	37.3	38.2	38.8	33.9	34.4	35.3	36.3	37.3	38.2	38.8
2	95th	104	105	105	106	107	107	108	57	58	58	59	60	61	61
	Height (in)	36.4	37	37.9	39	40.1	41.1	41.7	36.4	37	37.9	39	40.1	41.1	41.7
3	95th	106	106	107	107	108	109	109	60	61	61	62	63	64	64
	Height (in)	38.8	39.4	40.5	41.7	42.9	43.9	44.5	38.8	39.4	40.5	41.7	42.9	43.9	44.5
4	95th	107	107	108	108	109	110	110	63	64	65	66	67	67	68
	Height (in)	41.1	41.8	43.0	44.3	45.5	46.7	47.4	41.1	41.8	43.0	44.3	45.5	46.7	47.4
5	95th	107	108	109	109	110	111	112	66	67	68	69	70	70	71
	Height (in)	43.4	44.2	45.4	46.8	48.2	49.4	50.2	43.4	44.2	45.4	46.8	48.2	49.4	50.2
6	95th	108	109	110	111	112	113	114	69	70	70	71	72	72	73
	Height (in)	45.7	46.5	47.8	49.3	50.8	52.1	52.9	45.7	46.5	47.8	49.3	50.8	52.1	52.9
7	95th	110	110	111	112	114	115	116	71	71	72	73	73	74	74
	Height (in)	47.8	48.6	50	51.6	53.2	54.6	55.5	47.8	48.6	50	51.6	53.2	54.6	55.5
8	95th	111	112	112	114	115	116	117	72	73	73	74	75	75	75
	Height (in)	49.6	50.5	52	53.7	55.4	56.9	57.9	49.6	50.5	52	53.7	55.4	56.9	57.9
9	95th	112	112	113	115	116	118	119	74	74	75	76	76	77	77
	Height (in)	51.3	52.2	53.8	55.6	57.4	59.1	60.1	51.3	52.2	53.8	55.6	57.4	59.1	60.1
10	95th	112	113	114	116	118	120	121	76	76	77	77	78	78	78
	Height (in)	53	54	55.7	57.6	59.6	61.3	62.4	53	54	55.7	57.6	59.6	61.3	62.4
11	95th	114	114	116	118	120	123	124	77	78	78	78	78	78	78
	Height (in)	55.2	56.3	58.1	60.1	62.2	64	65.2	55.2	56.3	58.1	60.1	62.2	64	65.2
12	95th	116	117	118	121	124	126	128	78	78	78	78	78	79	79

Aim: To standardize the evaluation and management of patients with IgA vasculitis

Note 4: Follow-up recommendations- *most follow-up can be done by patient's PCP*

- **All follow-up appointments should include:** physical exam and weight (e.g. evaluate for peripheral edema), blood pressure measurement and urinalysis. Urine protein:Cr ratio should be calculated if 2+ protein identified on urinalysis. Should also include renal panel IF there is any history of renal involvement or if there are abnormal BP or urinalysis findings.
- If no kidney involvement:
    - Follow-up weekly for 4 weeks
    - THEN monthly until 6 months after diagnosis
- If renal dysfunction, hematuria or proteinuria (at initial presentation or later-onset), have patient avoid NSAIDs, consider Nephrology consult/referral (see note 7) and arrange follow-up – *Nephrology can help determine which of these follow-ups will be with Nephrology vs PCP*
    - Follow-up weekly for 4 weeks
    - THEN monthly for 5 months (until 6 months after diagnosis)
    - THEN every 3 months for an additional 6 months (until 12 months after diagnosis)
    - THEN every 6 months for a minimum of 4 years (until 5 years after diagnosis)
    - THEN yearly lifelong ongoing monitoring of blood pressure and UA in patients with a history of IgA vasculitis nephritis (i.e. hematuria + proteinuria)
- If patients are started on systemic steroids (see notes 5-6), they should be referred to follow up with Pediatric Endocrinology 1 month into the steroid course to assess for adrenal insufficiency

***If a patient started on steroids has a relapse after systemic steroids are weaned, the patient should be referred to rheumatology*

Aim: To standardize the evaluation and management of patients with IgA vasculitis

Note 5: Medications

Medication	Route Options	Dosing	Notes
Pain Medications – *uncomplicated abdominal pain and joint pains usually improve within 72 hours*			
Acetaminophen	Oral, Enteral, Rectal, IV	10-15 mg/kg/dose every 6-8 hours PRN Max 650 mg/dose	OK to use in combination with an NSAID.
Celecoxib (NSAID)	Oral, Enteral	Children: 1 mg/kg/dose every 12 hours PRN Adolescents: 100-200 mg/dose every 12 hours Max 200 mg/dose	NSAID of choice if patient has kidney involvement or a GI bleed
Ibuprofen (NSAID)	Oral, Enteral	5-10 mg/kg/dose every 6-8 hours PRN Max 600 mg/dose	Only use if patient has no nephritis or AKI and no active GI bleed.
Ketorolac (NSAID)	IV, IM	0.5 mg/kg/dose every 6-8 hours PRN Max 15 mg/dose	Only use if patient has no kidney involvement and no active GI bleed.
Naproxen (NSAID)	Oral, Enteral	5-7 mg/kg/dose every 8-12 hours PRN Max 1000 mg/**day**	Only use if patient has no kidney involvement and no active GI bleed.
Adjunct Pain Medications			
Hyoscyamine	Oral, Enteral	0.0625 – 0.125 mg every 4 hours PRN	Anti-cholinergic, anti-spasmodic used for pain related to cramping. Contraindicated if any concern for obstruction.
Methylprednisolone for debilitating pain* -See Note 6	IV	2 mg/kg/**day**, divided every 6-12 hours Max 60 mg/**day**	Give in addition to IV analgesics for severe abdominal and/or joint pain (or testicular pain)* Transition to oral prednisone when adequate PO intake (see wean below)
Prednisone/Prednisolone for debilitating pain* -See Note 6	Oral Liquid: Prednisolone Tablet: Prednisone	2 mg/kg/**day**, divided every 12-24 hours Max 60mg/**day**	Give in addition to IV analgesics for severe abdominal and/or joint pain (or testicular pain)* Wean Schedule (20% wean of original dose weekly): 1) 2 mg/kg/**day** divided BID for 5 days; (*Can start with 1 mg/kg/day divided BID, with a wean by 20% Q5 days, for milder symptoms) 2) 1.6 mg/kg/**day** divided BID for 5 days; 3) 1.2 mg/kg/**day** divided BID for 5 days; 4) 0.8 mg/kg/day once daily for 5 days; 5) 0.4 mg/kg/day once daily for 5 days; then discontinue. *Patients on steroids should be referred to follow-up with outpatient Endocrinology during this fifth week to assess for adrenal insufficiency. If symptoms recur or worsen during the wean, refer patient to Rheumatology Use with famotidine 0.5 mg/kg orally twice daily (max dose 20 mg)

** severe abdominal pain defined as pain that interferes with ability to eat, drink, sleep or do activities of daily living; severe joint pain defined as pain that interferes with ability to ambulate or do activities of daily living*

Aim: To standardize the evaluation and management of patients with IgA vasculitis

Note 6: Utility of steroids for patients with IgA vasculitis

- While steroids may reduce the duration of pain associated with IgA vasculitis, steroids do NOT alter long-term clinical outcomes in patients and should NOT be started empirically with the intention to prevent nephritis
- Some patients require steroids to treat IgA vasculitis nephritis or another complication (e.g. orchitis, cerebral vasculitis). In these cases, dosing and taper plan may be different than the regimen in note 5 and should be determined under the guidance of specialty consultants.
- Steroids should be used with caution in patients with intussusception as a complication of IgA vasculitis as there is a risk of obscuring the signs of recurrence

Note 7: Consult Nephrology in a patient with suspected IgA vasculitis if:

- Abnormal creatinine for age (inpatient nephrology consult or outpatient within 4 weeks)
- Macroscopic hematuria (inpatient nephrology consult or outpatient appointment within 4 weeks)
- Persistent microscopic hematuria for >6 months (outpatient referral)
- Persistent proteinuria (Pr:Cr ratio >0.5 mg/mg x 4 weeks) (inpatient nephrology consult or outpatient within 4 weeks)
- Nephrotic-range proteinuria (spot urine protein:creatinine ratio ≥ 2 mg/mg) (inpatient nephrology consult or call nephrologist on call for **urgent** appointment)
- Oliguria (urine output <0.5 ml/kg/hr x 24 hours) (inpatient nephrology consult)
- Hypertension (see note 3) (inpatient nephrology consult or call nephrologist on call for guidance)

*Nephrology may suggest a renal biopsy if the patient has progressive renal impairment, severe or persistent proteinuria, or nephritic syndrome

Note 8: IgA vasculitis and anticipatory guidance

- Renal manifestations generally present within the first 2 months but can take up to 6 months
- A first episode of IgA vasculitis, in the absence of significant renal diseases, usually resolves spontaneously within 4-6 weeks
- Around 1/3 of patients will have relapses/recurrence of symptoms, more likely in patients with renal disease.

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