Table of Contents:

Α.	Arthralgias	pg. 2
Β.	Joint Swelling, Joint Contracture, Limp Joint	pg. 2
C.	Weakness	pg. 3
D.	Back Pain	pg. 3
Ε.	Malar Rash	pg. 4
F.	Proteinuria and/or Hematuria	pg. 4
G.	Unexplained Fevers or Weight Loss	pg. 5
Η.	Skin Tightening or Extremity Color Changes	pg. 5
I.	Iritis	pg. 6
J.	Chronic Pain	pg. 6
Κ.	Positive (+) ANA	pg. 6

* These guidelines are to be used only as a tool for initial reference and not be used as exclusive indicators for referral to Rheumatology.

For appointments, please call the Patient Access Center at 888-770-2462 (888-770-CHOC) Fax ALL pertinent medical records to 855-246-2329 (855-CHOC-FAX) To speak with a CHOC Children's Specialist in Rheumatology, please call 714-509-8617 Website: <u>http://www.choc.org/specialists/rheumatology</u>

1 | P a g e September 29, 2015

A. Arthralgias [ICD-9 Code: 719.4*] [ICD-10 Code: M25.5*]

 Possible Diagnosis Systemic Juvenile Idiopathic Arthritis (JIA) Polyarticular Oligoarticular 	 Pre-Referral Evaluation Check for presence of: Joint swelling Hypermobility Flat feet 	 When to refer to Rheumatology If patient has persistent joint swelling, persistent limp or joint contracture If assistance is needed for managing hypermobility 	 Pre-Referral Workup CBC, Panel 18, ESR, UA, ANA, RF, HLA-B27 X-rays if appropriate 		
B.Joint Swelling, Joint	B.Joint Swelling, Joint Contracture, Limp Joint [ICD-9 Code: 719.0*] [ICD-10 Code: M25.4*]				
 Possible Diagnosis Systemic Juvenile Idiopathic Arthritis (JIA) Polyarticular Oligoarticular 	 Pre-Referral Evaluation Rule out infection, septic joint - if suspicious, refer urgently to Orthopaedics Document joint swelling, contractures Check X-rays as appropriate 	 When to refer to Rheumatology If patient has persistent joint swelling, persistent limp or joint contracture, not attributable to an Orthopedic problem 	 Pre-Referral Workup CBC, Panel 18, ESR, UA, ANA, RF, HLA-B27 X-rays if appropriate Place PPD 		

C. Weakness [ICD-9 Code: 728.87] [ICD-10 Code: M62.81]

Possible Diagnosis • Juvenile Dermatomyositis	 Pre-Referral Evaluation Check for proximal muscle weakness Check for presence of typical DM rash Consider MRI (w/o Gd) of prox muscles 	 When to refer to Rheumatology If weakness persists and is not attributable to a neurologic problem If there is a typical DM rash (Gotton's or Heliotrope) If the MRI shows muscle edema consistent with inflammation 	 Pre-Referral Workup ► CBC, Panel 18, ESR, UA, CPK, Aldolase ► Place PPD
D. Back Pain [ICD-9 Code:	724.5] [ICD-10 Code: M54.9]		
 Possible Diagnosis Juvenile Ankylosing Spondylitis (JAS) 	 Pre-Referral Evaluation Check for Sacroiliac Joint tenderness Check for ability to flex and extend back Consider MRI (w/Gd) of LS spine and SI joints 	 When to refer to Rheumatology If patient shows signs of SI joint tenderness or +MRI c/w inflammatory arthritis in SI joints/spine If there is significant decreased ROM in the back 	 Pre-Referral Workup CBC, Panel 18, ESR UA, HLA-B27 Place PPD CXR/spine films + SL jts

E. Malar Rash [ICD-9 Code: 782.1] [ICD-10 Code: R21]

Possible Diagnosis	Pre-Referral Evaluation	When to refer to Rheumatology	Pre-Referral Workup
Systemic Lupus Erythematosus (SLE)	 Monitor if rash persists over time, or becomes purpuric or eroded Check screening ANA and if positive, send full Lupus panel Check for other signs of Systemic Lupus 	 If rash persists or becomes purpuric or eroded If Lupus antibodies are positive (not just ANA) If patient has any other systemic signs of Lupus, including: joint swelling, oral ulcers, proteinuria, serositis, cytopenias, or mental status changes 	 CBC, Panel 18, ESR UA, HLA-B27 Place PPD CXR/spine films + SL jts
		status changes	

F. Proteinuria [ICD-9 Code: 791.0] [ICD-10: R80.9] Hematuria [ICD-9 Code: 599.70] [ICD-10 Code: R31.9]

Possible Diagnosis	Pre-Referral Evaluation	When to refer to Rheumatology	Pre-Referral Workup
 Systemic Lupus Erythematosus (SLE) ANCA - associated Vasculitis Goodpasture's Syndrome 	 Monitor if rash persists over time, or becomes purpuric or eroded Check 1st am urine for Prot/Creatinine Rule out infection (urine cx, Chlamydia/GC) Refer to <u>Nephrology Referral</u> <u>Guidelines</u> 	 If proteinuria persists and there is no infectious or anatomic cause found If lupus antibodies are positive If there are any other signs of systemic disease including: rash, fever, weight loss, arthritis, serositis, etc. 	 CBC, Panel 18, ESR, UA, HLA-B27 Place PPD CXR/spine films + SI jts

For appointments, please call the Patient Access Center at 888-770-2462 (888-770-CHOC) Fax ALL pertinent medical records to 855-246-2329 (855-CHOC-FAX) To speak with a CHOC Children's Specialist in Rheumatology, please call 714-509-8617 Website: <u>http://www.choc.org/specialists/rheumatology</u>

G. Unexplained Fevers [ICD-9 Code: 780.60] [ICD-10 Code: R50.9] Weight Loss [ICD-9 Code: 783.21] [ICD-10 Code: : R63.4]

Possible Diagnosis

- Systemic Juvenile Idiopathic Arthritis (JIA)
- Periodic Fevers Syndromes

Pre-Referral Evaluation

- Rule out infection first (Infectious Disease consult)
- Rule out malignancy (Oncology consult)
- Examine for signs of systemic autoimmune disease, especially arthritis

When to refer to Rheumatology

- If no evidence of infection or malignancy
- If there are specific signs of systemic disease - including: rash, oral ulcers, arthritis, serositis, etc.
- If there is a family history of periodic fevers

Pre-Referral Workup

- CBC, Panel 18, ESR, UA, ANA, Lupus Panel
- ► CXR
- ► Place PPD

H. Skin Tightening or Extremity Color Changes [ICD-9 Code: 709.8] [ICD-10 Code: : L98.8]

Possible Diagnosis

- Raynaud's Phenomenon
- Scleroderma

Pre-Referral Evaluation

 Examine for signs of sclerodactyly or skin tightening, esophageal dysmotility, calcinosis, pulmonary hypertension

When to refer to Rheumatology

- ► If there are progressive skin changes (inc. linear)
- ► If there are joint contractures
- If there are any signs of systemic disease

Pre-Referral Workup

- CBC, Panel 18, ESR, UA, ANA, Lupus Panel
- ► CXR
- ► Place PPD

I. Iritis [ICD-9 Code: 364.*] [ICD-10	Code: H20.0*]		
 Possible Diagnosis Juvenile Idiopathic Arthritis (JIA) Sarcoid ANCA Vasculitis 	 Pre-Referral Evaluation Refer urgently to Ophthalmology Examine for signs of systemic disease, especially arthritis 	 When to refer to Rheumatology If the Ophthalmologist confirms Uveitis and there is not an infectious cause found 	 Pre-Referral WorkuP ► CBC, Panel 18, ESR, UA, ► Urine Prot/Cr ► Place PPD
J. Chronic Pain [ICD-9 Code: 338	.29] [ICD-10 Code: G89.29]		
 Possible Diagnosis Fibromyalgia K. Positive (+) ANA [ICD-9 Code 	 Pre-Referral Evaluation Examine for specific source (joint swelling) Refer 1st to specific specialists (Neuro for headaches, GI for abdominal pain, etc.) 795 791 [ICD-10 Code: R76 0] 	 When to refer to Rheumatology Only if there is a specific source of pain, i.e. arthritis or myositis or there is lab evidence of inflammation (abn ESR) Refer to pain management and/or PT for Fibromyalgia 	 Pre-Referral Workup CBD, Panel 18, ESR, UA X-rays if appropriate
 Possible Diagnosis Pauci JIA SLE Hashimoto's 	 Pre-Referral Evaluation Examine for specific of autoimmune disease (jt swelling, rash, etc.) 	 When to refer to Rheumatology If patient has specific clinical signs to autoimmune disease (not just a (+) ANA 	 Pre-Referral Workup ► CBC, Panel 18, ESR, UA ► T4, TSH ► Lupus Panel

For appointments, please call the Patient Access Center at 888-770-2462 (888-770-CHOC) Fax ALL pertinent medical records to 855-246-2329 (855-CHOC-FAX) To speak with a CHOC Children's Specialist in Rheumatology, please call 714-509-8617 Website: <u>http://www.choc.org/specialists/rheumatology</u>

Sources used in development of these Referral Guidelines:

Jennifer E. Weiss MD, Norman T. Ilowite MD Juvenile Idiopathic Arthritis Rheumatic Disease Clinics of North America Volume 33, Issue 3, August 2007, Pages 441-470

Boon S, McCurdy D, "Systemic Lupus Erythematosus in Children", Pediatric Annals, Vol.31 no.7 August 2002 (Page 407 - 425)

Lucy R. Wedderburn, Lisa G. Rider. Juvenile dermatomyositis: new developments inpathogenesis, assessment and treatment. Best Practice & Research Clinical Rheumatology. 23 (2009) 665-678.

Stichweh, Dorotheea; Arce, Edsela; Pascual, Virginia Update on Pediatric Systemic Lupus Erythematosus Current Opinion in Rheumatology: September 2004 - Volume 16 - Issue 5 - pp 577-587

Villa-Forte, A.: European League Against Rheumatism: European Vasculitis Study Group: European League Againty Rheumatism/European Vasculitis Study Group Recommendations for the Management of Vasculitis, Curr Opin Rheumaol 2010: 22: 49-53

Boon, S.: Pediatric Hospital Medicine, Textbook of Inpatient Management, Chapters 70 ("Systemic Lupus Erythemtosus"), 72 & 73 ("Unusual Rheumatologic & Vasculitic Diseases"; Editors: Perkin ,Swift, Newton & Anas; Lipponcott Williams & Wilkins June 2008

Textbook of Pediatric Rheumatology (Fifth Edition) Copyright © 2005 Elsevier Inc. All rights reserved Edited by: James T. Cassidy, MD, and Ross E. Petty, MD, PhD

For appointments, please call the Patient Access Center at 888-770-2462 (888-770-CHOC) Fax ALL pertinent medical records to 855-246-2329 (855-CHOC-FAX) To speak with a CHOC Children's Specialist in Rheumatology, please call 714-509-8617 Website: <u>http://www.choc.org/specialists/rheumatology</u>

7 | P a g e September 29, 2015