

# Tocilizumab: Actemra®; Tofidence™ (Intravenous)

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## I. Length of Authorization <sup>1,2,18,30</sup>

Coverage will be provided as follows:

- Castleman Disease: 4 months and may be renewed
- Cytokine Release Syndrome: 4 doses only and may NOT be renewed
- Immune Checkpoint Inhibitor Related Toxicities: 1 dose only\* and may NOT be renewed
- Giant Cell Arteritis: 1 year of therapy
- All other indications: 6 months and may be renewed

## II. Dosing Limits

### A. Quantity Limit (max daily dose) [NDC Unit]:

- Actemra 80 mg/4 mL single-dose vial: 1 vial per 14 days
- Actemra 200 mg/10 mL single-dose vial: 3 vials per 28 days
- Actemra 400 mg/20 mL single-dose vial: 2 vials per 14 days
- Tofidence 80 mg/4 mL single-dose vial: 1 vial per 14 days
- Tofidence 200 mg/10 mL single-dose vial: 3 vials per 28 days
- Tofidence 400 mg/20 mL single-dose vial: 2 vials per 14 days

### B. Max Units (per dose and over time) [HCPCS Unit]:

Diagnosis	Billable Units	Interval (days)
Giant Cell Arteritis	600	28
Rheumatoid Arthritis & Polyarticular Juvenile Idiopathic Arthritis, NMOSD	800	28
Systemic Juvenile Idiopathic Arthritis, Castleman's Disease (B-Cell Lymphomas) & Acute Graft Versus Host Disease (aGVHD)	800	14

Cytokine Release Syndrome (CRS)	3200	1 course of therapy only
Immune Checkpoint Inhibitor Related Toxicities	800	1 course of therapy only

### III. Initial Approval Criteria <sup>1,2</sup>

Coverage is provided in the following conditions:

Site of care specialty infusion program requirements are met (refer to [Moda Site of Care Policy](#)).

- Self-administered injectable medications are not covered when supplied in a provider's office, clinic or facility.

- Patient is at least 18 years of age (unless otherwise specified); **AND**

#### Universal Criteria <sup>1,2</sup>

- Patient has been evaluated and screened for the presence of latent tuberculosis (TB) infection prior to initiating treatment and will receive ongoing monitoring for presence of TB during treatment; **AND**
- Patient does not have an active infection, including clinically important localized infections; **AND**
- Patient will not receive live vaccines during therapy; **AND**
- Patient is not on concurrent treatment with another IL-inhibitor, TNF-inhibitor, biologic response modifier or other non-biologic agent (i.e., abrocitinib, apremilast, tofacitinib, baricitinib, upadacitinib, deucravacitinib, etc.), unless otherwise specified; **AND**

#### Rheumatoid Arthritis † <sup>1,2,20,28</sup>

- Physician has assessed baseline disease severity utilizing an objective measure/tool; **AND**
- Documented moderate to severe active disease; **AND**
- Patient has had at least a 3 month trial and failed previous therapy with ONE oral disease modifying anti-rheumatic agent (DMARD) such as methotrexate, azathioprine, auranofin, hydroxychloroquine, penicillamine, sulfasalazine, leflunomide, etc.; **AND**
- May be used as a single agent or in combination with methotrexate or other non-biologic DMARDs (e.g., methotrexate, hydroxychloroquine, leflunomide, sulfasalazine, etc.)

- Patient must try and have an inadequate response, contraindication, or intolerance to at least a three (3) month trial of Enbrel **AND** Humira; **OR**

- Patient is continuing treatment

#### Giant Cell Arteritis (GCA) † <sup>1,30-33</sup>

- Patient has large vessel arteritis that has at some point been verified with biopsy or with imaging of the large vessels (color Doppler ultrasound [CDUS], MRI, PET-CT, or CT angiography); **AND**
- Patient has active disease and an elevated c-reactive protein (CRP) and erythrocyte sedimentation rate (ESR); **AND**
- Patient has had an inadequate response, contraindication, or intolerance to glucocorticoid therapy alone; **AND**
- Used in combination with a tapering course of glucocorticoids (*NOTE: Actemra can be used alone following discontinuation of glucocorticoids.*)

**Juvenile Idiopathic Arthritis (JIA) † Φ<sup>1,2,6,19</sup>**

- Patient is at least 2 years of age; **AND**
- Physician has assessed baseline disease severity utilizing an objective measure/tool; **AND**
- Patient has active systemic juvenile idiopathic arthritis (sJIA) OR polyarticular juvenile idiopathic arthritis (pJIA); **AND**
- Patient has had at least a 1-month trial and failure (unless contraindicated or intolerant) of previous therapy with either oral non-steroidal anti-inflammatory drugs (NSAIDs) OR an oral disease-modifying anti-rheumatic agent (DMARD) (e.g., methotrexate, leflunomide, sulfasalazine, etc.); **AND**
- May be used as a single agent or in combination with methotrexate

*(Note: The following applies to polyarticular (PJIA) disease only)*

- Patient must try and have an inadequate response, contraindication, or intolerance to at least a three (3) month trial of Enbrel AND Humira; **OR**
- Patient is continuing treatment

**Castleman Disease (B-Cell Lymphomas) ‡<sup>3</sup>**

- Used as a single agent; **AND**
  - Patient has unicentric disease; **AND**
    - Patient is human immunodeficiency virus (HIV)-negative and human herpesvirus-8 (HHV-8)-negative; **AND**
    - Used as second-line therapy for relapsed or refractory disease; **OR**
  - Patient has multicentric disease; **AND**
    - Used as subsequent therapy for relapsed, refractory, or progressive disease

**Cytokine Release Syndrome (CRS) † Φ<sup>1,3</sup>**

- Patient is at least 2 years of age; **AND**
  - Patient has received or will be receiving chimeric antigen receptor (CAR) T cell therapy (e.g., axicabtagene ciloleucel, brexucabtagene autoleucel, idecabtagene vicleucel, lisocabtagene maraleucel, tisagenlecleucel, ciltacabtagene autoleucel, etc.); **AND**

- Tocilizumab is being ordered to have on-hand, prior to the administration of CAR-T therapy, if needed for the treatment of CRS; **OR**
- Used for management of one of the following:
  - Prolonged (>3 days) G1 CRS in patients with significant symptoms, comorbidities, and/or in elderly patients; **OR**
  - G1 CRS symptoms that persist >24 hours in patients who have been treated with axicabtagene ciloleucel or brexucabtagene autoleucel; **OR**
  - G1 CRS that develops <72 hours after infusion in patients who have been treated with lisocabtagene maraleucel; **OR**
  - G2-4 CRS; **OR**
  - G1-4 neurotoxicity as additional single-dose therapy if the patient has concurrent CRS; **OR**
- Used as supportive care in patients with acute lymphoblastic leukemia (ALL) who have severe CRS related to blinatumomab therapy

#### **Management of Immune Checkpoint Inhibitor Related Toxicities ‡ 3,18,27**

- Patient has been receiving therapy with an immune checkpoint inhibitor (e.g. nivolumab, pembrolizumab, atezolizumab, avelumab, durvalumab, cemiplimab, ipilimumab, dostarlimab nivolumab/relatlimab-rmbw, retifanlimab, etc.); **AND**
  - Used as additional therapy for management of immunotherapy-related giant cell arteritis; **OR**
  - Used as additional disease modifying antirheumatic therapy (DMARD) for moderate or severe immunotherapy-related inflammatory arthritis; **AND**
    - Patient's symptoms have not improved after holding immunotherapy; **AND**
      - Patient has not responded to oral corticosteroids; **OR**
      - Patient is unable to taper corticosteroids; **OR**
  - Patient has polymyalgia rheumatica and is unable to taper prednisone OR has no improvement in symptoms

#### **Acute Graft Versus Host Disease (aGVHD) ‡ 3,21-23,29**

- Patient has received an allogeneic hematopoietic stem cell transplant; **AND**
- Used for steroid-refractory acute GVHD; **AND**
- Used in combination with systemic corticosteroids as additional therapy following no response to first-line therapies

#### **Neuromyelitis Optica Spectrum Disorder (NMOSD) ‡ 24-26,34**

- Patient has a confirmed diagnosis based on the following:
  - Patient is seropositive for aquaporin-4 (AQP4) IgG antibodies; **AND**

- Patient has at least one core clinical characteristic § (*\*Note: some core clinical characteristics require both clinical and typical MRI findings*); **AND**
- Alternative diagnoses have been excluded (e.g., myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOGAD), multiple sclerosis, sarcoidosis, cancer, chronic infection, etc.); **OR**
- Patient is seronegative for AQP-4 IgG antibodies **OR** has unknown AQP-4-IgG status; **AND**
  - Patient has at least two core clinical characteristics§ occurring as a result of one or more clinical attacks; **AND**
  - Patient experienced **ALL** of the following:
    - At least 1 core clinical characteristic must be acute optic neuritis, acute myelitis with LETM\*, or area postrema syndrome; **AND**
    - Fulfillment of typical MRI finding requirements for each area affected **ψ**; **AND**
  - Alternative diagnoses have been excluded (e.g., myelin oligodendrocyte glycoprotein (MOG) antibody disease (MOGAD), multiple sclerosis, sarcoidosis, cancer, chronic infection, etc.); **AND**
- Used as a single agent or in combination with immunosuppressive therapy (e.g. azathioprine, methotrexate, mycophenolate, etc.)

§ Core Clinical Characteristics of NMOSD <sup>24,34</sup>
<ul style="list-style-type: none"> <li>▪ Acute optic neuritis</li> <li>▪ Acute myelitis</li> <li>▪ Acute area postrema syndrome (APS): episode of otherwise unexplained hiccups and/or nausea and vomiting (lasting for at least 48 hours or with MRI evidence of a dorsal brainstem lesion)</li> <li>▪ Acute brainstem syndrome other than APS</li> <li>▪ Symptomatic narcolepsy or acute diencephalic clinical syndrome with NMOSD-typical diencephalic MRI lesions <b>∕</b></li> <li>▪ Acute cerebral syndrome with NMOSD-typical brain lesions on MRI <b>§</b></li> </ul>
ψ Typical MRI findings in NMOSD related to clinical presentation (T2 unless noted otherwise) <sup>34</sup>
<ul style="list-style-type: none"> <li>▪ Optic neuritis: Normal cerebral MRI (or only nonspecific white matter lesions) <b>OR</b> longitudinally extensive optic nerve lesion (≥ half of the length of the optic nerve or involving optic chiasm; T2 or T1/Gd)</li> <li>▪ Myelitis: Intramedullary lesion ≥ 3 contiguous VS (LETM) <b>OR</b> focal atrophy ≥ 3 contiguous VS in patients with a history of acute myelitis</li> <li>▪ Area postrema syndrome (APS): Lesion in the dorsal medulla oblongata/area postrema</li> <li>▪ Other brainstem syndrome: Periependymal brainstem lesion (4th ventricle)</li> <li>▪ <b>∕</b> Diencephalic syndrome: Periependymal lesion (3rd ventricle) <b>OR</b> hypothalamic/thalamic lesion</li> <li>▪ <b>§</b> Cerebral syndrome: Extensive periependymal lesion (lateral ventricle; often with Gd) <b>OR</b> long (&gt; 1/2 length), diffuse, heterogeneous or edematous corpus callosum lesion <b>OR</b> long corticospinal tract lesion (unilateral or bilateral, contiguously involving internal capsule and cerebral peduncle) <b>OR</b> large, confluent (unilateral or bilateral) subcortical or deep white matter lesion</li> </ul>
*LETM = longitudinally extensive transverse myelitis lesions

† FDA Approved Indication(s); ‡ Compendia Recommended Indication(s); Ⓞ Orphan Drug

## IV. Renewal Criteria <sup>1,2</sup>

Coverage can be renewed based upon the following criteria:

- Patient continues to meet the universal and other indication-specific relevant criteria identified in section III; **AND**
- Absence of unacceptable toxicity from the drug. Examples of unacceptable toxicity include: serious infections, severe neutropenia, severe thrombocytopenia, severe hepatotoxicity, gastrointestinal perforation, immunosuppression, severe hypersensitivity reactions, demyelinating disorders, etc.; **AND**

### **Oncology Indications**

#### **Castleman Disease (B-Cell Lymphomas) <sup>3</sup>**

- Disease response with treatment as defined by stabilization of disease or decrease in size of tumor or tumor spread

#### **Acute Graft Versus Host Disease (aGVHD) <sup>21-23,29</sup>**

- Response to therapy with an improvement in one or more of the following:
  - Clinician assessments (e.g., NIH Skin Score, Upper GI Response Score, NIH Lung Symptom Score, etc.)
  - Patient-reported symptoms (e.g., Lee Symptom Scale, etc.)

### **Non-Oncology Indications**

#### **Rheumatoid arthritis (RA) <sup>13-15</sup>**

- Disease response as indicated by improvement in signs and symptoms compared to baseline such as the number of tender and swollen joint counts, reduction of C-reactive protein, improvement of patient global assessment, and/or an improvement on a disease activity scoring tool [e.g. an improvement on a composite scoring index such as Disease Activity Score-28 (DAS28) of 1.2 points or more or a  $\geq 20\%$  improvement on the American College of Rheumatology-20 (ACR20) criteria]

#### **Giant Cell Arteritis <sup>1,30-32</sup>**

- Disease response as indicated by improvement in signs and compared to baseline such as headache, temporal artery tenderness, visual symptoms, inflammatory parameters (e.g., erythrocyte sedimentation rate [ESR], C-reactive protein), etc.

#### **Juvenile Idiopathic Arthritis (sJIA/pJIA) <sup>16,17</sup>**

- Disease response as indicated by improvement in signs and symptoms compared to baseline such as the number of tender and swollen joint counts, reduction of C-reactive protein, improvement of patient global assessment, and/or an improvement on a disease activity scoring tool [e.g., an improvement on a composite scoring index such as Juvenile Arthritis

Disease Activity Score (JADAS) or the American College of Rheumatology (ACR) Pediatric (ACR-Pedi 30) of at least 30% improvement from baseline in three of six variables].

**Cytokine Release Syndrome** <sup>1,3</sup>

- May not be renewed

**Management of Immune Checkpoint Inhibitor Related Toxicities** <sup>18</sup>

- May not be renewed

**NMOSD** <sup>24,25</sup>

- Disease response as indicated by stabilization/improvement in any of the following:
  - Neurologic symptoms as evidenced by a decrease in acute relapses or improvement of stability
  - Reduced hospitalizations
  - Reduction/discontinuation in plasma exchange treatments
  - Reduction/discontinuation of corticosteroids without relapse

**V. Dosage/Administration** <sup>1,2,9,18,21-23,26,29</sup>

Doses exceeding 800 mg per infusion are not recommended (unless otherwise specified).

Indication	Dose
Adult Rheumatoid Arthritis	Administer 4 mg/kg intravenously every 4 weeks <ul style="list-style-type: none"> <li>• May increase to 8 mg/kg every 4 weeks based on clinical response, up to a maximum of 800 mg per dose.</li> </ul>
Polyarticular Juvenile Idiopathic Arthritis	<u>Weight ≥ 30 kg</u> <ul style="list-style-type: none"> <li>• Administer 8 mg/kg intravenously every 4 weeks</li> </ul> <u>Weight &lt; 30 kg</u> <ul style="list-style-type: none"> <li>• Administer 10 mg/kg intravenously every 4 weeks</li> </ul>
Systemic Juvenile Idiopathic Arthritis	<u>Weight ≥ 30 kg</u> <ul style="list-style-type: none"> <li>• Administer 8 mg/kg intravenously every 2 weeks</li> </ul> <u>Weight &lt; 30 kg</u> <ul style="list-style-type: none"> <li>• Administer 12 mg/kg intravenously every 2 weeks</li> </ul>
Castleman Disease (B-Cell Lymphomas)	Administer 8 mg/kg intravenously every 2 weeks for 16 weeks (8 doses total)
Cytokine Release Syndrome (CRS)	<u>Weight ≥ 30 kg</u> <ul style="list-style-type: none"> <li>• Administer 8 mg/kg intravenously every 8 hours, if needed, up to a maximum of 4 total doses*</li> </ul> <u>Weight &lt; 30 kg</u>

Indication	Dose
	<ul style="list-style-type: none"> <li>Administer 12 mg/kg intravenously every 8 hours, if needed, up to a maximum of 4 total doses*</li> </ul> <p><i>*If no clinical improvement in the signs and symptoms of CRS occurs after the first dose, up to 3 additional doses may be administered. The interval between consecutive doses should be at least 8 hours. May be used with or without corticosteroids. Doses exceeding 800 mg per infusion are not recommended in CRS patients.</i></p>
Immune-Checkpoint Inhibitor Related Toxicities	<p>Administer 4 mg/kg intravenously one time only*</p> <p><i>*Additional doses may be approved on a case by case basis</i></p>
Acute GVHD	Administer 8 mg/kg intravenously, every 2-4 weeks until disease progression or unacceptable toxicity
NMOSD	Administer 8 mg/kg intravenously, every 4 weeks
Giant Cell Arteritis	<p>Administer 6 mg/kg intravenously every 4 weeks in combination with a tapering course of glucocorticoids</p> <p><i>*Doses exceeding 600 mg per infusion are not recommended</i></p> <p><i>**Actemra can be used alone following discontinuation of glucocorticoids.</i></p>
<b>Note:</b> Only Actemra IV is FDA approved for Giant Cell Arteritis (GCA) and Cytokine Release Syndrome (CRS)	

## VI. Billing Code/Availability Information

### HCPCS Code:

- J3262 – Injection, tocilizumab, 1 mg; 1 billable unit = 1 mg (*Actemra IV Only*)
- Q5133 – Injection, tocilizumab-bavi (tofidence), biosimilar, 1 mg; 1 billable unit = 1 mg (*Tofidence Only*) (*Effective 04/01/2024*)
- J3590 – Unclassified biologics (*Tofidence Only*) (*Discontinue use on 04/01/2024*)

### NDC(s):

- Actemra 80 mg/4 mL single-dose vial: 50242-0135-xx
- Actemra 200 mg/10 mL single-dose vial: 50242-0136-xx
- Actemra 400 mg/20 mL single-dose vial: 50242-0137-xx
- Tofidence 80 mg/4 mL single-dose vial: 64406-0024-xx
- Tofidence 200 mg/10 mL single-dose vial: 64406-0022-xx
- Tofidence 400 mg/20 mL single-dose vial: 64406-0023-xx

## VII. References

- Actemra [package insert]. South San Francisco, CA; Genentech, Inc.; December 2022. Accessed September 2023.



2. Tofidence [package insert]. Cambridge, MA; Biogen MA Inc.; September 2023. Accessed October 2023.
3. Referenced with permission from the NCCN Drugs & Biologics Compendium (NCCN Compendium®) tocilizumab. National Comprehensive Cancer Network, 2023. The NCCN Compendium® is a derivative work of the NCCN Guidelines®. NATIONAL COMPREHENSIVE CANCER NETWORK®, NCCN®, and NCCN GUIDELINES® are trademarks owned by the National Comprehensive Cancer Network, Inc.” To view the most recent and complete version of the Compendium, go online to NCCN.org. Accessed September 2023.
4. Singh JA, Saag KG, Bridges SL Jr, et al. 2015 American College of Rheumatology Guideline for the Treatment of Rheumatoid Arthritis. *Arthritis Care Res (Hoboken)*. 2015 Nov 6. doi: 10.1002/acr.22783.
5. Beukelman T, Patkar NM, Saag KG, et al. 2011 American College of Rheumatology recommendations for the treatment of juvenile idiopathic arthritis: initiation and safety monitoring of therapeutic agents for the treatment of arthritis and systemic features. *Arthritis Care Res (Hoboken)*. 2011 Apr;63(4):465-82.
6. Ringold S, Weiss PF, Beukelman T, et al. 2013 update of the 2011 American College of Rheumatology recommendations for the treatment of juvenile idiopathic arthritis: recommendations for the medical therapy of children with systemic juvenile idiopathic arthritis and tuberculosis screening among children receiving biologic medications. *Arthritis Rheum*. 2013 Oct;65(10):2499-512.
7. Ringold, S, Angeles-Han, S, Beukelman, T, et al. 2019 American College of Rheumatology/Arthritis Foundation Guideline for the Treatment of Juvenile Idiopathic Arthritis: Therapeutic Approaches for Non-Systemic Polyarthritis, Sacroiliitis, and Enthesitis. *Arthritis Care Res*, 71: 717-734.
8. DeWitt EM, Kimura Y, Beukelman T, et al. Consensus treatment plans for new-onset systemic juvenile idiopathic arthritis. *Arthritis Care Res (Hoboken)*. 2012 Jul;64(7):1001-10.
9. Nishimoto N, Kanakura Y, Aozasa K, et al. Humanized anti-interleukin-6 receptor antibody treatment of multicentric Castleman disease. *Blood* 2005;106:2627-2632
10. Smolen JS, Landewé R, Bijlsma J, et al. EULAR recommendations for the management of rheumatoid arthritis with synthetic and biological disease-modifying antirheumatic drugs: 2016 update. *Ann Rheum Dis*. 2017 Mar 6. pii: annrheumdis-2016-210715.
11. Fraser JA, Weyand CM, Newman NJ, Biousse V. The treatment of giant cell arteritis. *Rev Neurol Dis*. 2008 Summer;5(3):140-52.
12. Dasgupta B, Borg FA, Hassan N, et al. BSR and BHRP guidelines for the management of giant cell arteritis. *Rheumatology (Oxford)*. 2010 Aug;49(8):1594-7.
13. National Institute for Health and Care Excellence. NICE 2018. Rheumatoid Arthritis in Adults: Management. Published 11 July 2018. Last updated 12 October 2020. NICE Guideline [NG100]. <https://www.nice.org.uk/guidance/ng100>. Accessed September 2023.

14. National Institute for Health and Care Excellence. NICE 2010. Adalimumab, etanercept, infliximab, rituximab and abatacept for the treatment of rheumatoid arthritis after failure of a TNF inhibitor. Published 25 August 2010. Technology appraisal guidance [TA195]. <https://www.nice.org.uk/guidance/ta195>. Accessed September 2023.
15. Ward MM, Guthri LC, Alba MI. Rheumatoid Arthritis Response Criteria And Patient-Reported Improvement in Arthritis Activity: Is an ACR20 Response Meaningful to Patients”. *Arthritis Rheumatol*. 2014 Sep; 66(9): 2339–2343. doi: 10.1002/art.38705
16. Ringold S, Bittner R, Neggi T, et al. Performance of rheumatoid arthritis disease activity measures and juvenile arthritis disease activity scores in polyarticular-course juvenile idiopathic arthritis: Analysis of their ability to classify the American College of Rheumatology pediatric measures of response and the preliminary criteria for flare and inactive disease. *Arthritis Care Res (Hoboken)*. 2010 Aug;62(8):1095-102.
17. Consolaro A, Giancane G, Schiappapietra B, et al. Clinical outcome measures in juvenile idiopathic arthritis. *Pediatric Rheumatology* 18 April 2016 14:23.
18. Stroud C, Hedge A, Cherry C, et al. Tocilizumab for the management of immune mediated adverse events secondary to PD-1 blockage. *Journal of Oncology Pharmacy Practice*. 2017 December. <https://doi.org/10.1177/1078155217745144>.
19. Ringold S, Angeles-Han ST, Beukelman T, et al. 2019 American College of Rheumatology/Arthritis Foundation Guideline for the Treatment of Juvenile Idiopathic Arthritis: Therapeutic Approaches for Non-Systemic Polyarthritis, Sacroiliitis, and Entesitis. *Arthritis Care & Research*, Vol. 71, No. 6, June 2019, pp 717–734 DOI 10.1002/acr.23870.
20. Smolen JS, Landewé RBM, Bijlsma JWJ, et al. EULAR recommendations for the management of rheumatoid arthritis with synthetic and biological disease-modifying antirheumatic drugs: 2019 update. *Annals of the Rheumatic Diseases* 2020;79:685-699.
21. Yucebay F, Matthews C, Puto M, et al. Tocilizumab as first-line therapy for steroid-refractory acute graft-versus-host-disease: analysis of a single-center experience, *Leukemia & Lymphoma*. 2019 60:9, 2223-2229, DOI: 10.1080/10428194.2019.1573996
22. Ganetsky A, Frey NV, Hexner EO, et al. Tocilizumab for the treatment of severe steroid-refractory acute graft-versus-host disease of the lower gastrointestinal tract. *Bone Marrow Transplant*. 54, 212–217 (2019). <https://doi.org/10.1038/s41409-018-0236-z>
23. Dobryski WR, Pasquini M, Kovatovic K, et al. Tocilizumab for the Treatment of Steroid Refractory Graft-versus-Host Disease. 17:12, 1862-1868 (2011). DOI:<https://doi.org/10.1016/j.bbmt.2011.07.001>
24. Wingerchuk DM, Banwell B, Bennett JL, et al. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. *Neurology*. 2015 Jul;85(2):177-89. Epub 2015 Jun 19.
25. Trebst C, Jarius S, Berthele A, et al. Update on the diagnosis and treatment of neuromyelitis optica: recommendations of the Neuromyelitis Optica Study Group (NEMOS). *J Neurol* 2014; 261:1.

26. Zhang C, Zhang M, Qiu W, et al, TANGO Study Investigators. Safety and efficacy of tocilizumab versus azathioprine in highly relapsing neuromyelitis optica spectrum disorder (TANGO): an open-label, multicentre, randomised, phase 2 trial. *Lancet Neurol*. 2020;19(5):391
27. Stone J, Tuckwell K, Dimonaco S, et al. Trial of Tocilizumab in Giant-Cell Arteritis. *N Engl J Med* 2017; 377:317-328 doi: 10.1056/NEJMoa1613849
28. Fraenkel L, Bathon JM, England BR, et al. 2021 American College of Rheumatology Guideline for the Treatment of Rheumatoid Arthritis. *Arthritis Rheumatol*. 2021 Jul;73(7):1108-1123. doi: 10.1002/art.41752.
29. Roddy JV, Haverkos BM, McBride A, et al. Tocilizumab for steroid refractory acute graft-versus-host disease. *Leuk Lymphoma*. 2016;57(1):81-5. doi: 10.3109/10428194.2015.1045896.
30. Turesson C, Börjesson O, Larsson K, et al. (2019) Swedish Society of Rheumatology 2018 guidelines for investigation, treatment, and follow-up of giant cell arteritis, *Scandinavian Journal of Rheumatology*, 48:4, 259-265, doi: 10.1080/03009742.2019.1571223.
31. Mackie SL, Dejaco C, Appenzeller S, et al. British Society for Rheumatology guideline on diagnosis and treatment of giant cell arteritis. *Rheumatology (Oxford)*. 2020 Mar 1;59(3):e1-e23. doi: 10.1093/rheumatology/kez672.
32. Ehlers L, Askling J, Bijlsma HW, et al. 2018 EULAR recommendations for a core data set to support observational research and clinical care in giant cell arteritis. *Annals of the Rheumatic Diseases* 2019;78:1160-1166.
33. Maz M, Chung SA, Abril A, et. al. 2021 American College of Rheumatology/Vasculitis Foundation Guideline for the Management of Giant Cell Arteritis and Takayasu Arteritis. *Arthritis Rheumatol*. 2021 Aug;73(8):1349-1365. doi: 10.1002/art.41774. Epub 2021 Jul 8. PMID: 34235884.
34. Jarius, S., Aktas, O., Ayzenberg, I. et al. Update on the diagnosis and treatment of neuromyelitis optica spectrum disorders (NMOSD) – revised recommendations of the Neuromyelitis Optica Study Group (NEMOS). Part I: Diagnosis and differential diagnosis. *J Neurol* 270, 3341–3368 (2023). <https://doi.org/10.1007/s00415-023-11634-0>

## Appendix 1 – Covered Diagnosis Codes

ICD-10	ICD-10 Description
C83.50	Lymphoblastic (diffuse) lymphoma unspecified site
C83.51	Lymphoblastic (diffuse) lymphoma lymph nodes of head, face, and neck
C83.52	Lymphoblastic (diffuse) lymphoma intrathoracic lymph nodes
C83.53	Lymphoblastic (diffuse) lymphoma intra-abdominal lymph nodes
C83.54	Lymphoblastic (diffuse) lymphoma lymph nodes of axilla and upper limb
C83.55	Lymphoblastic (diffuse) lymphoma lymph nodes of inguinal region and lower limb
C83.56	Lymphoblastic (diffuse) lymphoma intrapelvic lymph nodes

ICD-10	ICD-10 Description
C83.57	Lymphoblastic (diffuse) lymphoma spleen
C83.58	Lymphoblastic (diffuse) lymphoma lymph nodes of multiple sites
C83.59	Lymphoblastic (diffuse) lymphoma extranodal and solid organ sites
C91.00	Acute lymphoblastic leukemia not having achieved remission
C91.01	Acute lymphoblastic leukemia, in remission
C91.02	Acute lymphoblastic leukemia, in relapse
D47.Z2	Castleman disease
D89.810	Acute graft-versus-host disease
D89.812	Acute on chronic graft-versus-host disease
D89.813	Graft-versus-host disease, unspecified
D89.831	Cytokine release syndrome, grade 1
D89.832	Cytokine release syndrome, grade 2
D89.833	Cytokine release syndrome, grade 3
D89.834	Cytokine release syndrome, grade 4
D89.839	Cytokine release syndrome, grade unspecified
G36.0	Neuromyelitis optica [Devic]
G92.00	Immune effector cell-associated neurotoxicity syndrome, grade unspecified
G92.01	Immune effector cell-associated neurotoxicity syndrome, grade 1
G92.02	Immune effector cell-associated neurotoxicity syndrome, grade 2
G92.03	Immune effector cell-associated neurotoxicity syndrome, grade 3
G92.04	Immune effector cell-associated neurotoxicity syndrome, grade 4
M05.10	Rheumatoid lung disease with rheumatoid arthritis of unspecified site
M05.111	Rheumatoid lung disease with rheumatoid arthritis of right shoulder
M05.112	Rheumatoid lung disease with rheumatoid arthritis of left shoulder
M05.119	Rheumatoid lung disease with rheumatoid arthritis of unspecified shoulder
M05.121	Rheumatoid lung disease with rheumatoid arthritis of right elbow
M05.122	Rheumatoid lung disease with rheumatoid arthritis of left elbow
M05.129	Rheumatoid lung disease with rheumatoid arthritis of unspecified elbow
M05.131	Rheumatoid lung disease with rheumatoid arthritis of right wrist
M05.132	Rheumatoid lung disease with rheumatoid arthritis of left wrist
M05.139	Rheumatoid lung disease with rheumatoid arthritis of unspecified wrist
M05.141	Rheumatoid lung disease with rheumatoid arthritis of right hand
M05.142	Rheumatoid lung disease with rheumatoid arthritis of left hand
M05.149	Rheumatoid lung disease with rheumatoid arthritis of unspecified hand
M05.151	Rheumatoid lung disease with rheumatoid arthritis of right hip
M05.152	Rheumatoid lung disease with rheumatoid arthritis of left hip
M05.159	Rheumatoid lung disease with rheumatoid arthritis of unspecified hip
M05.161	Rheumatoid lung disease with rheumatoid arthritis of right knee

ICD-10	ICD-10 Description
M05.162	Rheumatoid lung disease with rheumatoid arthritis of left knee
M05.169	Rheumatoid lung disease with rheumatoid arthritis of unspecified knee
M05.171	Rheumatoid lung disease with rheumatoid arthritis of right ankle and foot
M05.172	Rheumatoid lung disease with rheumatoid arthritis of left ankle and foot
M05.179	Rheumatoid lung disease with rheumatoid arthritis of unspecified ankle and foot
M05.19	Rheumatoid lung disease with rheumatoid arthritis of multiple sites
M05.20	Rheumatoid vasculitis with rheumatoid arthritis of unspecified site
M05.211	Rheumatoid vasculitis with rheumatoid arthritis of right shoulder
M05.212	Rheumatoid vasculitis with rheumatoid arthritis of left shoulder
M05.219	Rheumatoid vasculitis with rheumatoid arthritis of unspecified shoulder
M05.221	Rheumatoid vasculitis with rheumatoid arthritis of right elbow
M05.222	Rheumatoid vasculitis with rheumatoid arthritis of left elbow
M05.229	Rheumatoid vasculitis with rheumatoid arthritis of unspecified elbow
M05.231	Rheumatoid vasculitis with rheumatoid arthritis of right wrist
M05.232	Rheumatoid vasculitis with rheumatoid arthritis of left wrist
M05.239	Rheumatoid vasculitis with rheumatoid arthritis of unspecified wrist
M05.241	Rheumatoid vasculitis with rheumatoid arthritis of right hand
M05.242	Rheumatoid vasculitis with rheumatoid arthritis of left hand
M05.249	Rheumatoid vasculitis with rheumatoid arthritis of unspecified hand
M05.251	Rheumatoid vasculitis with rheumatoid arthritis of right hip
M05.252	Rheumatoid vasculitis with rheumatoid arthritis of left hip
M05.259	Rheumatoid vasculitis with rheumatoid arthritis of unspecified hip
M05.261	Rheumatoid vasculitis with rheumatoid arthritis of right knee
M05.262	Rheumatoid vasculitis with rheumatoid arthritis of left knee
M05.269	Rheumatoid vasculitis with rheumatoid arthritis of unspecified knee
M05.271	Rheumatoid vasculitis with rheumatoid arthritis of right ankle and foot
M05.272	Rheumatoid vasculitis with rheumatoid arthritis of left ankle and foot
M05.279	Rheumatoid vasculitis with rheumatoid arthritis of unspecified ankle and foot
M05.29	Rheumatoid vasculitis with rheumatoid arthritis of multiple sites
M05.30	Rheumatoid heart disease with rheumatoid arthritis of unspecified site
M05.311	Rheumatoid heart disease with rheumatoid arthritis of right shoulder
M05.312	Rheumatoid heart disease with rheumatoid arthritis of left shoulder
M05.319	Rheumatoid heart disease with rheumatoid arthritis of unspecified shoulder
M05.321	Rheumatoid heart disease with rheumatoid arthritis of right elbow
M05.322	Rheumatoid heart disease with rheumatoid arthritis of left elbow
M05.329	Rheumatoid heart disease with rheumatoid arthritis of unspecified elbow
M05.331	Rheumatoid heart disease with rheumatoid arthritis of right wrist
M05.332	Rheumatoid heart disease with rheumatoid arthritis of left wrist

ICD-10	ICD-10 Description
M05.339	Rheumatoid heart disease with rheumatoid arthritis of unspecified wrist
M05.341	Rheumatoid heart disease with rheumatoid arthritis of right hand
M05.342	Rheumatoid heart disease with rheumatoid arthritis of left hand
M05.349	Rheumatoid heart disease with rheumatoid arthritis of unspecified hand
M05.351	Rheumatoid heart disease with rheumatoid arthritis of right hip
M05.352	Rheumatoid heart disease with rheumatoid arthritis of left hip
M05.359	Rheumatoid heart disease with rheumatoid arthritis of unspecified hip
M05.361	Rheumatoid heart disease with rheumatoid arthritis of right knee
M05.362	Rheumatoid heart disease with rheumatoid arthritis of left knee
M05.369	Rheumatoid heart disease with rheumatoid arthritis of unspecified knee
M05.371	Rheumatoid heart disease with rheumatoid arthritis of right ankle and foot
M05.372	Rheumatoid heart disease with rheumatoid arthritis of left ankle and foot
M05.379	Rheumatoid heart disease with rheumatoid arthritis of unspecified ankle and foot
M05.39	Rheumatoid heart disease with rheumatoid arthritis of multiple sites
M05.40	Rheumatoid myopathy with rheumatoid arthritis of unspecified site
M05.411	Rheumatoid myopathy with rheumatoid arthritis of right shoulder
M05.412	Rheumatoid myopathy with rheumatoid arthritis of left shoulder
M05.419	Rheumatoid myopathy with rheumatoid arthritis of unspecified shoulder
M05.421	Rheumatoid myopathy with rheumatoid arthritis of right elbow
M05.422	Rheumatoid myopathy with rheumatoid arthritis of left elbow
M05.429	Rheumatoid myopathy with rheumatoid arthritis of unspecified elbow
M05.431	Rheumatoid myopathy with rheumatoid arthritis of right wrist
M05.432	Rheumatoid myopathy with rheumatoid arthritis of left wrist
M05.439	Rheumatoid myopathy with rheumatoid arthritis of unspecified wrist
M05.441	Rheumatoid myopathy with rheumatoid arthritis of right hand
M05.442	Rheumatoid myopathy with rheumatoid arthritis of left hand
M05.449	Rheumatoid myopathy with rheumatoid arthritis of unspecified hand
M05.451	Rheumatoid myopathy with rheumatoid arthritis of right hip
M05.452	Rheumatoid myopathy with rheumatoid arthritis of left hip
M05.459	Rheumatoid myopathy with rheumatoid arthritis of unspecified hip
M05.461	Rheumatoid myopathy with rheumatoid arthritis of right knee
M05.462	Rheumatoid myopathy with rheumatoid arthritis of left knee
M05.469	Rheumatoid myopathy with rheumatoid arthritis of unspecified knee
M05.471	Rheumatoid myopathy with rheumatoid arthritis of right ankle and foot
M05.472	Rheumatoid myopathy with rheumatoid arthritis of left ankle and foot
M05.479	Rheumatoid myopathy with rheumatoid arthritis of unspecified ankle and foot
M05.49	Rheumatoid myopathy with rheumatoid arthritis of multiple sites
M05.50	Rheumatoid myopathy with rheumatoid arthritis of unspecified sites

ICD-10	ICD-10 Description
M05.511	Rheumatoid polyneuropathy with rheumatoid arthritis of right shoulder
M05.512	Rheumatoid polyneuropathy with rheumatoid arthritis of left shoulder
M05.519	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified shoulder
M05.521	Rheumatoid polyneuropathy with rheumatoid arthritis of right elbow
M05.522	Rheumatoid polyneuropathy with rheumatoid arthritis of left elbow
M05.529	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified elbow
M05.531	Rheumatoid polyneuropathy with rheumatoid arthritis of right wrist
M05.532	Rheumatoid polyneuropathy with rheumatoid arthritis of left wrist
M05.539	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified wrist
M05.541	Rheumatoid polyneuropathy with rheumatoid arthritis of right hand
M05.542	Rheumatoid polyneuropathy with rheumatoid arthritis of left hand
M05.549	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified hand
M05.551	Rheumatoid polyneuropathy with rheumatoid arthritis of right hip
M05.552	Rheumatoid polyneuropathy with rheumatoid arthritis of left hip
M05.559	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified hip
M05.561	Rheumatoid polyneuropathy with rheumatoid arthritis of right knee
M05.562	Rheumatoid polyneuropathy with rheumatoid arthritis of left knee
M05.569	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified knee
M05.571	Rheumatoid polyneuropathy with rheumatoid arthritis of right ankle and foot
M05.572	Rheumatoid polyneuropathy with rheumatoid arthritis of left ankle and foot
M05.579	Rheumatoid polyneuropathy with rheumatoid arthritis of unspecified ankle and foot
M05.59	Rheumatoid polyneuropathy with rheumatoid arthritis of multiple sites
M05.60	Rheumatoid arthritis of unspecified site with involvement of other organs and systems
M05.611	Rheumatoid arthritis of right shoulder with involvement of other organs and systems
M05.612	Rheumatoid arthritis of left shoulder with involvement of other organs and systems
M05.619	Rheumatoid arthritis of unspecified shoulder with involvement of other organs and systems
M05.621	Rheumatoid arthritis of right elbow with involvement of other organs and systems
M05.622	Rheumatoid arthritis of left elbow with involvement of other organs and systems
M05.629	Rheumatoid arthritis of unspecified elbow with involvement of other organs and systems
M05.631	Rheumatoid arthritis of right wrist with involvement of other organs and systems
M05.632	Rheumatoid arthritis of left wrist with involvement of other organs and systems
M05.639	Rheumatoid arthritis of unspecified wrist with involvement of other organs and systems
M05.641	Rheumatoid arthritis of right hand with involvement of other organs and systems
M05.642	Rheumatoid arthritis of left hand with involvement of other organs and systems
M05.649	Rheumatoid arthritis of unspecified hand with involvement of other organs and systems
M05.651	Rheumatoid arthritis of right hip with involvement of other organs and systems
M05.652	Rheumatoid arthritis of left hip with involvement of other organs and systems
M05.659	Rheumatoid arthritis of unspecified hip with involvement of other organs and systems

ICD-10	ICD-10 Description
M05.661	Rheumatoid arthritis of right knee with involvement of other organs and systems
M05.662	Rheumatoid arthritis of left knee with involvement of other organs and systems
M05.669	Rheumatoid arthritis of unspecified knee with involvement of other organs and systems
M05.671	Rheumatoid arthritis of right ankle and foot with involvement of other organs and systems
M05.672	Rheumatoid arthritis of left ankle and foot with involvement of other organs and systems
M05.679	Rheumatoid arthritis of unspecified ankle and foot with involvement of other organs and systems
M05.69	Rheumatoid arthritis of multiple sites with involvement of other organs and systems
M05.7A	Rheumatoid arthritis with rheumatoid factor of other specified site without organ or systems
M05.711	Rheumatoid arthritis with rheumatoid factor of right shoulder without organ or systems involvement
M05.712	Rheumatoid arthritis with rheumatoid factor of left shoulder without organ or systems involvement
M05.719	Rheumatoid arthritis with rheumatoid factor of unspecified shoulder without organ or systems
M05.721	Rheumatoid arthritis with rheumatoid factor of right elbow without organ or systems involvement
M05.722	Rheumatoid arthritis with rheumatoid factor of left elbow without organ or systems involvement
M05.729	Rheumatoid arthritis with rheumatoid factor of unspecified elbow without organ or systems
M05.731	Rheumatoid arthritis with rheumatoid factor of right wrist without organ or systems involvement
M05.732	Rheumatoid arthritis with rheumatoid factor of left wrist without organ or systems involvement
M05.739	Rheumatoid arthritis with rheumatoid factor of unspecified wrist without organ or systems
M05.741	Rheumatoid arthritis with rheumatoid factor of right hand without organ or systems involvement
M05.742	Rheumatoid arthritis with rheumatoid factor of left hand without organ or systems involvement
M05.749	Rheumatoid arthritis with rheumatoid factor of unspecified hand without organ or systems
M05.751	Rheumatoid arthritis with rheumatoid factor of right hip without organ or systems involvement
M05.752	Rheumatoid arthritis with rheumatoid factor of left hip without organ or systems involvement
M05.759	Rheumatoid arthritis with rheumatoid factor of unspecified hip without organ or systems involvement
M05.761	Rheumatoid arthritis with rheumatoid factor of right knee without organ or systems involvement
M05.762	Rheumatoid arthritis with rheumatoid factor of left knee without organ or systems involvement
M05.769	Rheumatoid arthritis with rheumatoid factor of unspecified knee without organ or systems
M05.771	Rheumatoid arthritis with rheumatoid factor of right ankle and foot without organ or systems
M05.772	Rheumatoid arthritis with rheumatoid factor of left ankle and foot without organ or systems
M05.779	Rheumatoid arthritis with rheumatoid factor of unspecified ankle and wrist without organ or systems
M05.79	Rheumatoid arthritis with rheumatoid factor of multiple sites without organ or systems involvement
M05.8A	Other rheumatoid arthritis with rheumatoid factor of other specified site
M05.811	Other rheumatoid arthritis with rheumatoid factor of right shoulder
M05.812	Other rheumatoid arthritis with rheumatoid factor of left shoulder
M05.819	Other rheumatoid arthritis with rheumatoid factor of unspecified shoulder
M05.821	Other rheumatoid arthritis with rheumatoid factor of right elbow
M05.822	Other rheumatoid arthritis with rheumatoid factor of left elbow
M05.829	Other rheumatoid arthritis with rheumatoid factor of unspecified elbow
M05.831	Other rheumatoid arthritis with rheumatoid factor of right wrist



ICD-10	ICD-10 Description
M05.832	Other rheumatoid arthritis with rheumatoid factor of left wrist
M05.839	Other rheumatoid arthritis with rheumatoid factor of unspecified wrist
M05.841	Other rheumatoid arthritis with rheumatoid factor of right hand
M05.842	Other rheumatoid arthritis with rheumatoid factor of left hand
M05.849	Other rheumatoid arthritis with rheumatoid factor of unspecified hand
M05.851	Other rheumatoid arthritis with rheumatoid factor of right hip
M05.852	Other rheumatoid arthritis with rheumatoid factor of left hip
M05.859	Other rheumatoid arthritis with rheumatoid factor of unspecified hip
M05.861	Other rheumatoid arthritis with rheumatoid factor of right knee
M05.862	Other rheumatoid arthritis with rheumatoid factor of left knee
M05.869	Other rheumatoid arthritis with rheumatoid factor of unspecified knee
M05.871	Other rheumatoid arthritis with rheumatoid factor of right ankle and foot
M05.872	Other rheumatoid arthritis with rheumatoid factor of left ankle and foot
M05.879	Other rheumatoid arthritis with rheumatoid factor of unspecified ankle and foot
M05.89	Other rheumatoid arthritis with rheumatoid factor of multiple sites
M05.9	Rheumatoid arthritis with rheumatoid factor, unspecified
M06.0A	Rheumatoid arthritis without rheumatoid factor, other specified site
M06.019	Rheumatoid arthritis without rheumatoid factor, unspecified shoulder
M06.021	Rheumatoid arthritis without rheumatoid factor, right elbow
M06.022	Rheumatoid arthritis without rheumatoid factor, left elbow
M06.029	Rheumatoid arthritis without rheumatoid factor, unspecified elbow
M06.031	Rheumatoid arthritis without rheumatoid factor, right wrist
M06.032	Rheumatoid arthritis without rheumatoid factor, left wrist
M06.039	Rheumatoid arthritis without rheumatoid factor, unspecified wrist
M06.041	Rheumatoid arthritis without rheumatoid factor, right hand
M06.042	Rheumatoid arthritis without rheumatoid factor, left hand
M06.049	Rheumatoid arthritis without rheumatoid factor, unspecified hand
M06.051	Rheumatoid arthritis without rheumatoid factor, right hip
M06.052	Rheumatoid arthritis without rheumatoid factor, left hip
M06.059	Rheumatoid arthritis without rheumatoid factor, unspecified hip
M06.061	Rheumatoid arthritis without rheumatoid factor, right knee
M06.062	Rheumatoid arthritis without rheumatoid factor, left knee
M06.069	Rheumatoid arthritis without rheumatoid factor, unspecified knee
M06.071	Rheumatoid arthritis without rheumatoid factor, right ankle and foot
M06.072	Rheumatoid arthritis without rheumatoid factor, left ankle and foot
M06.079	Rheumatoid arthritis without rheumatoid factor, unspecified ankle and foot
M06.08	Rheumatoid arthritis without rheumatoid factor, vertebrae
M06.09	Rheumatoid arthritis without rheumatoid factor, multiple sites

ICD-10	ICD-10 Description
M06.4	Inflammatory polyarthropathy
M06.8A	Other specified rheumatoid arthritis, other specified site
M06.811	Other specified rheumatoid arthritis, right shoulder
M06.812	Other specified rheumatoid arthritis, left shoulder
M06.819	Other specified rheumatoid arthritis, unspecified shoulder
M06.821	Other specified rheumatoid arthritis, right elbow
M06.822	Other specified rheumatoid arthritis, left elbow
M06.829	Other specified rheumatoid arthritis, unspecified elbow
M06.831	Other specified rheumatoid arthritis, right wrist
M06.832	Other specified rheumatoid arthritis, left wrist
M06.839	Other specified rheumatoid arthritis, unspecified wrist
M06.841	Other specified rheumatoid arthritis, right hand
M06.842	Other specified rheumatoid arthritis, left hand
M06.849	Other specified rheumatoid arthritis, unspecified hand
M06.851	Other specified rheumatoid arthritis, right hip
M06.852	Other specified rheumatoid arthritis, left hip
M06.859	Other specified rheumatoid arthritis, unspecified hip
M06.861	Other specified rheumatoid arthritis, right knee
M06.862	Other specified rheumatoid arthritis, left knee
M06.869	Other specified rheumatoid arthritis, unspecified knee
M06.871	Other specified rheumatoid arthritis, right ankle and foot
M06.872	Other specified rheumatoid arthritis, left ankle and foot
M06.879	Other specified rheumatoid arthritis, unspecified ankle and foot
M06.88	Other specified rheumatoid arthritis, vertebrae
M06.89	Other specified rheumatoid arthritis, multiple sites
M06.9	Rheumatoid arthritis, unspecified
M08.0A	Unspecified juvenile rheumatoid arthritis, other specified site
M08.011	Unspecified juvenile rheumatoid arthritis, right shoulder
M08.012	Unspecified juvenile rheumatoid arthritis, left shoulder
M08.019	Unspecified juvenile rheumatoid arthritis, unspecified shoulder
M08.021	Unspecified juvenile rheumatoid arthritis, right elbow
M08.022	Unspecified juvenile rheumatoid arthritis, left elbow
M08.029	Unspecified juvenile rheumatoid arthritis, unspecified elbow
M08.031	Unspecified juvenile rheumatoid arthritis, right wrist
M08.032	Unspecified juvenile rheumatoid arthritis, left wrist
M08.039	Unspecified juvenile rheumatoid arthritis, unspecified wrist
M08.041	Unspecified juvenile rheumatoid arthritis, right hand
M08.042	Unspecified juvenile rheumatoid arthritis, left hand

ICD-10	ICD-10 Description
M08.049	Unspecified juvenile rheumatoid arthritis, unspecified hand
M08.051	Unspecified juvenile rheumatoid arthritis, right hip
M08.052	Unspecified juvenile rheumatoid arthritis, left hip
M08.059	Unspecified juvenile rheumatoid arthritis, unspecified hip
M08.061	Unspecified juvenile rheumatoid arthritis, right knee
M08.062	Unspecified juvenile rheumatoid arthritis, left knee
M08.069	Unspecified juvenile rheumatoid arthritis, unspecified knee
M08.071	Unspecified juvenile rheumatoid arthritis, right ankle and foot
M08.072	Unspecified juvenile rheumatoid arthritis, left ankle and foot
M08.079	Unspecified juvenile rheumatoid arthritis, unspecified ankle and foot
M08.08	Unspecified juvenile rheumatoid arthritis, vertebrae
M08.09	Unspecified juvenile rheumatoid arthritis, multiple sites
M08.2A	Juvenile rheumatoid arthritis with systemic onset, other specified site
M08.211	Juvenile rheumatoid arthritis with systemic onset, right shoulder
M08.212	Juvenile rheumatoid arthritis with systemic onset, left shoulder
M08.219	Juvenile rheumatoid arthritis with systemic onset, unspecified shoulder
M08.221	Juvenile rheumatoid arthritis with systemic onset, right elbow
M08.222	Juvenile rheumatoid arthritis with systemic onset, left elbow
M08.229	Juvenile rheumatoid arthritis with systemic onset, unspecified elbow
M08.231	Juvenile rheumatoid arthritis with systemic onset, right wrist
M08.232	Juvenile rheumatoid arthritis with systemic onset, left wrist
M08.239	Juvenile rheumatoid arthritis with systemic onset, unspecified wrist
M08.241	Juvenile rheumatoid arthritis with systemic onset, right hand
M08.242	Juvenile rheumatoid arthritis with systemic onset, left hand
M08.249	Juvenile rheumatoid arthritis with systemic onset, unspecified hand
M08.251	Juvenile rheumatoid arthritis with systemic onset, right hip
M08.252	Juvenile rheumatoid arthritis with systemic onset, left hip
M08.259	Juvenile rheumatoid arthritis with systemic onset, unspecified hip
M08.261	Juvenile rheumatoid arthritis with systemic onset, right knee
M08.262	Juvenile rheumatoid arthritis with systemic onset, left knee
M08.269	Juvenile rheumatoid arthritis with systemic onset, unspecified knee
M08.271	Juvenile rheumatoid arthritis with systemic onset, right ankle and foot
M08.272	Juvenile rheumatoid arthritis with systemic onset, left ankle and foot
M08.279	Juvenile rheumatoid arthritis with systemic onset, unspecified ankle and foot
M08.28	Juvenile rheumatoid arthritis with systemic onset, vertebrae
M08.29	Juvenile rheumatoid arthritis with systemic onset, multiple sites
M08.3	Juvenile rheumatoid polyarthritis (seronegative)
M08.4A	Pauciarticular juvenile rheumatoid arthritis, other specified site

ICD-10	ICD-10 Description
M08.411	Pauciarticular juvenile rheumatoid arthritis, right shoulder
M08.412	Pauciarticular juvenile rheumatoid arthritis, left shoulder
M08.419	Pauciarticular juvenile rheumatoid arthritis, unspecified shoulder
M08.421	Pauciarticular juvenile rheumatoid arthritis, right elbow
M08.422	Pauciarticular juvenile rheumatoid arthritis, left elbow
M08.429	Pauciarticular juvenile rheumatoid arthritis, unspecified elbow
M08.431	Pauciarticular juvenile rheumatoid arthritis, right wrist
M08.432	Pauciarticular juvenile rheumatoid arthritis, left wrist
M08.439	Pauciarticular juvenile rheumatoid arthritis, unspecified wrist
M08.441	Pauciarticular juvenile rheumatoid arthritis, right hand
M08.442	Pauciarticular juvenile rheumatoid arthritis, left hand
M08.449	Pauciarticular juvenile rheumatoid arthritis, unspecified hand
M08.451	Pauciarticular juvenile rheumatoid arthritis, right hip
M08.452	Pauciarticular juvenile rheumatoid arthritis, left hip
M08.459	Pauciarticular juvenile rheumatoid arthritis, unspecified hip
M08.461	Pauciarticular juvenile rheumatoid arthritis, right knee
M08.462	Pauciarticular juvenile rheumatoid arthritis, left knee
M08.469	Pauciarticular juvenile rheumatoid arthritis, unspecified knee
M08.471	Pauciarticular juvenile rheumatoid arthritis, right ankle and foot
M08.472	Pauciarticular juvenile rheumatoid arthritis, left ankle and foot
M08.479	Pauciarticular juvenile rheumatoid arthritis, unspecified ankle and foot
M08.48	Pauciarticular juvenile rheumatoid arthritis, vertebrae
M08.80	Other juvenile arthritis, unspecified site
M08.811	Other juvenile arthritis, right shoulder
M08.812	Other juvenile arthritis, left shoulder
M08.819	Other juvenile arthritis, unspecified shoulder
M08.821	Other juvenile arthritis, right elbow
M08.822	Other juvenile arthritis, left elbow
M08.829	Other juvenile arthritis, unspecified elbow
M08.831	Other juvenile arthritis, right wrist
M08.832	Other juvenile arthritis, left wrist
M08.839	Other juvenile arthritis, unspecified wrist
M08.841	Other juvenile arthritis, right hand
M08.842	Other juvenile arthritis, left hand
M08.849	Other juvenile arthritis, unspecified hand
M08.851	Other juvenile arthritis, right hip
M08.852	Other juvenile arthritis, left hip
M08.859	Other juvenile arthritis, unspecified hip

ICD-10	ICD-10 Description
M08.861	Other juvenile arthritis, right knee
M08.862	Other juvenile arthritis, left knee
M08.869	Other juvenile arthritis, unspecified knee
M08.871	Other juvenile arthritis, right ankle and foot
M08.872	Other juvenile arthritis, left ankle and foot
M08.879	Other juvenile arthritis, unspecified ankle and foot
M08.88	Other juvenile arthritis, other specified site
M08.89	Other juvenile arthritis, multiple sites
M08.9A	Juvenile arthritis, unspecified, other specified site
M08.911	Juvenile arthritis, unspecified, right shoulder
M08.912	Juvenile arthritis, unspecified, left shoulder
M08.919	Juvenile arthritis, unspecified, unspecified shoulder
M08.921	Juvenile arthritis, unspecified, right elbow
M08.922	Juvenile arthritis, unspecified, left elbow
M08.929	Juvenile arthritis, unspecified, unspecified elbow
M08.931	Juvenile arthritis, unspecified, right wrist
M08.932	Juvenile arthritis, unspecified, left wrist
M08.939	Juvenile arthritis, unspecified, unspecified wrist
M08.941	Juvenile arthritis, unspecified, right hand
M08.942	Juvenile arthritis, unspecified, left hand
M08.949	Juvenile arthritis, unspecified, unspecified hand
M08.951	Juvenile arthritis, unspecified, right hip
M08.952	Juvenile arthritis, unspecified, left hip
M08.959	Juvenile arthritis, unspecified, unspecified hip
M08.961	Juvenile arthritis, unspecified, right knee
M08.962	Juvenile arthritis, unspecified, left knee
M08.969	Juvenile arthritis, unspecified, unspecified knee
M08.971	Juvenile arthritis, unspecified, right ankle and foot
M08.972	Juvenile arthritis, unspecified, left ankle and foot
M08.979	Juvenile arthritis, unspecified, unspecified ankle and foot
M08.98	Juvenile arthritis, unspecified, vertebrae
M08.99	Juvenile arthritis, unspecified, multiple sites
M31.5	Giant cell arteritis with polymyalgia rheumatica
M31.6	Other giant cell arteritis
M35.3	Polymyalgia rheumatica
T80.82XA	Complication of immune effector cellular therapy, initial encounter
T80.82XS	Complication of immune effector cellular therapy, sequela
T80.89XA	Other complications following infusion, transfusion and therapeutic injection, initial encounter

ICD-10	ICD-10 Description
T80.89XS	Other complications following infusion, transfusion and therapeutic injection, sequela
T86.09	Other complications of bone marrow transplant

## Appendix 2 – Centers for Medicare and Medicaid Services (CMS)

Medicare coverage for outpatient (Part B) drugs is outlined in the Medicare Benefit Policy Manual (Pub. 100-2), Chapter 15, §50 Drugs and Biologicals. In addition, National Coverage Determination (NCD), Local Coverage Determinations (LCDs), and Local Coverage Articles (LCAs) may exist and compliance with these policies is required where applicable. They can be found at:

<https://www.cms.gov/medicare-coverage-database/search.aspx>. Additional indications may be covered at the discretion of the health plan.

Medicare Part B Covered Diagnosis Codes (applicable to existing NCD/LCD/LCA): N/A

Medicare Part B Administrative Contractor (MAC) Jurisdictions		
Jurisdiction	Applicable State/US Territory	Contractor
E (1)	CA, HI, NV, AS, GU, CNMI	Noridian Healthcare Solutions, LLC
F (2 & 3)	AK, WA, OR, ID, ND, SD, MT, WY, UT, AZ	Noridian Healthcare Solutions, LLC
5	KS, NE, IA, MO	Wisconsin Physicians Service Insurance Corp (WPS)
6	MN, WI, IL	National Government Services, Inc. (NGS)
H (4 & 7)	LA, AR, MS, TX, OK, CO, NM	Novitas Solutions, Inc.
8	MI, IN	Wisconsin Physicians Service Insurance Corp (WPS)
N (9)	FL, PR, VI	First Coast Service Options, Inc.
J (10)	TN, GA, AL	Palmetto GBA, LLC
M (11)	NC, SC, WV, VA (excluding below)	Palmetto GBA, LLC
L (12)	DE, MD, PA, NJ, DC (includes Arlington & Fairfax counties and the city of Alexandria in VA)	Novitas Solutions, Inc.
K (13 & 14)	NY, CT, MA, RI, VT, ME, NH	National Government Services, Inc. (NGS)
15	KY, OH	CGS Administrators, LLC