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**MB&RB**

**CHANGE 182  
6010.57-M  
MAY 22, 2017**

**PUBLICATIONS SYSTEM CHANGE TRANSMITTAL FOR  
TRICARE POLICY MANUAL (TPM), FEBRUARY 2008**

The Defense Health Agency has authorized the following addition(s)/revision(s).

**CHANGE TITLE:** EVOLVING PRACTICES RARE DISEASES 17-001

**CONREQ:** 18550

**PAGE CHANGE(S):** See page 2.

**SUMMARY OF CHANGE(S):** See pages 3 and 4.

**EFFECTIVE DATE:** See pages 3 and 4.

**IMPLEMENTATION DATE:** June 22, 2017.

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**6010.57-M**  
**MAY 22, 2017**

**REMOVE PAGE(S)**

**CHAPTER 1**

Section 3.1, pages 1 through 4

**INSERT PAGE(S)**

Section 3.1, pages 1 through 5

## **SUMMARY OF CHANGES**

### **CHAPTER 1**

#### Chapter 1, Section 3.1.

1. This change confirms the off-label use of rituximab injections for the following treatments:
  - a. Recurrent nodular CD20 positive lymphocyte predominant Hodgkin's disease. EFFECTIVE DATE: January 1, 2003.
  - b. Stiff Person Syndrome. EFFECTIVE DATE: March 31, 2005.
  - c. Immunoglobulin A (IgA) nephropathy (proliferative glomerulonephritis) to reduce proteinuria. EFFECTIVE DATE: May 1, 2007.
  - d. Neuromyelitis optica. EFFECTIVE DATE: March 26, 2010.
  - e. N-methyl-D-aspartate (NMDA) receptor encephalitis. EFFECTIVE DATE: July 20, 2016.
  - f. Constitutional (pure) red blood cell aplasia. EFFECTIVE DATE: August 22, 2016.
  - g. Autoimmune sclerosing pancreatitis. EFFECTIVE DATE: September 16, 2016.
  - h. Immunoglobulin G4-Related Disease (IgG4-RD). EFFECTIVE DATE: October 6, 2016.
  - i. Autoimmune hemolytic anemia. EFFECTIVE DATE: October 27, 2016.
  - j. Graft-Versus-Host-Disease (GVHD). EFFECTIVE DATE: November 1, 2016.
  - k. Bullous pemphigoid. EFFECTIVE DATE: November 9, 2016.
  - l. As a second-line treatment for autoimmune encephalitis. EFFECTIVE DATE: November 14, 2016.
  - m. Cryoglobulinemia. EFFECTIVE DATE: November 22, 2016.
  - n. Thrombotic Thrombocytopenic Purpura (TTP). EFFECTIVE DATE: January 3, 2017.
  - o. Polymyositis. EFFECTIVE DATE: January 19, 2017.
2. Chemotherapy injections (CPT procedure code 96542) for the treatment of Central Giant Cell Granuloma (CGCG) of the mandible is covered. EFFECTIVE DATE: July 22, 2016.
3. Peg interferon alfa-2A/180 (HCPCS J3490) for the treatment of Central Giant Cell Granuloma (CGCG) of the mandible is covered. EFFECTIVE DATE: July 22, 2016.
4. Fluorodeoxyglucose (FDG) PET scan (CPT1 procedure code 78815) for the treatment of Takayasu's Arteritis (also known as aortic arch syndrome) is covered. EFFECTIVE DATE: August 11, 2016.
5. Gammagard liquid injection (HCPCS J1596) for the treatment of Branch retinal artery occlusion secondary to Susac's Syndrome is covered. EFFECTIVE DATE: August 22, 2016.

**SUMMARY OF CHANGES (Continued)**

**CHAPTER 1 (Continued)**

Chapter 1, Section 3.1 (Continued).

6. An autologous bone marrow transplant, the harvest of autologous stem cells, and the cryopreservation of stem cells for the treatment of recurrent medulloblastoma is covered. EFFECTIVE DATE: October 13, 2016.
7. Magnetic Resonance-guided High Intensity Focused Ultrasound (MRgFUS) for the treatment of Desmoid fibromatosis is covered. EFFECTIVE DATE: December 15, 2016.
8. Stereotactic Body Radiation Therapy (SBRT) (CPT procedure codes 77435 and 77373) for the treatment of a benign neoplasm of the aortic body and other paraganglia is covered. EFFECTIVE DATE: January 24, 2017.

## Chapter 1

## Section 3.1

### Rare Diseases

Issue Date: May 18, 1994

Authority: [32 CFR 199.2\(b\)](#) and [32 CFR 199.4\(g\)\(15\)\(ii\)](#)

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#### 1.0 DESCRIPTION

TRICARE defines a rare disease as any disease or condition that **has a prevalence of less than 200,000 persons in the United States.**

#### 2.0 POLICY

**2.1** Coverage for treatment of rare diseases may be considered on a case-by-case basis. **In reviewing the case, the contractor is authorized to approve coverage when it is determined that the proposed treatment for the rare disease is medically necessary, including that the treatment is safe and effective.**

**2.1.1** In reviewing the case, any or all of the following **sources of clinical literature** may be used to determine if the proposed **treatment** is considered safe and effective.

**2.1.1.1** Trials published in refereed medical literature.

**2.1.1.2** Formal technology assessments.

**2.1.1.3** National medical policy organization positions.

**2.1.1.4** National professional associations.

**2.1.1.5** National expert opinion organizations.

**2.2** **In those situations where the contractor finds the proposed treatment is not considered safe and effective, the contractor shall forward the case to the Medical Benefits & Reimbursement Section (MB&RS) to permit the Director, Defense Health Agency (DHA), or designee, to complete an individual case review and make a determination in accordance with [32 CFR 199.4\(g\)\(15\)\(ii\)](#).**

**2.3** **TRICARE Encounter Data (TED) Record Special Processing Code "RD - Rare Diseases" shall be coded on all TED records where the contractor has approved treatment for a rare disease. Assignment of Special Processing Code RD will allow the DHA to identify procedures approved by contractors under the Rare Diseases policy and will allow bypass of TED edit 2-160-01R when the procedure code is on the No Government Pay Procedure Code List.**

**2.4** The contractor shall provide a monthly report as described in the Contract Data Requirements List (CDRL). The report should not include the rare disease treatments previously approved for coverage and listed herein.

**2.5** Off-label use of rituximab may be considered for cost-sharing for the following treatments:

**2.5.1** Effective January 1, 2003, for recurrent nodular CD20 positive lymphocyte predominant Hodgkin's disease.

**2.5.2** Effective March 31, 2005, for Stiff Person Syndrome.

**2.5.3** Effective May 1, 2007, for Immunoglobulin A (IgA) nephropathy (proliferative glomerulonephritis) to reduce proteinuria.

**2.5.4** Effective March 26, 2010, for neuromyelitis optica.

**2.5.5** Effective July 20, 2016, for N-methyl-D-aspartate (NMDA) receptor encephalitis.

**2.5.6** Effective August 22, 2016, for constitutional (pure) red blood cell aplasia.

**2.5.7** Effective September 16, 2016, for autoimmune sclerosing pancreatitis.

**2.5.8** Effective October 6, 2016, Immunoglobulin G4-related disease (IgG4-RD).

**2.5.9** Effective October 27, 2016, for autoimmune hemolytic anemia.

**2.5.10** Effective November 1, 2016, for Graft-Versus-Host-Disease (GVHD).

**2.5.11** Effective November 9, 2016, for bullous pemphigoid.

**2.5.12** Effective November 14, 2016, as a second-line treatment for autoimmune encephalitis.

**2.5.13** Effective November 22, 2016, for cryoglobulinemia.

**2.5.14** Effective January 3, 2017, for Thrombotic Thrombocytopenic Purpura (TTP).

**2.5.15** Effective January 19, 2017, for polymyositis.

**2.6** Effective May 13, 2009, Intraperitoneal Hyperthermic Chemotherapy (IPHC) (Current Procedural Terminology (CPT)<sup>1</sup> procedure codes 77600, 77605, and 96445) in conjunction with cytoreductive surgery or peritonectomy for treatment of pseudomyxoma peritonei resulting from appendiceal carcinoma may be covered on a case-by-case basis for adult patients when all of the following criteria are met:

- There is no evidence of distant metastasis.
- There is evidence of low histological aggressiveness of the disease.

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- The patient has not undergone preoperative systemic chemotherapy.
- The patient's condition does not preclude major surgery.
- The chemotherapeutic agents used are Mitomycin C, Cisplatin (also known as Cisplatinum), or Fluorouracil.

**2.7** Effective January 21, 2009, External Infusion Pumps (EIPs) for insulin may be considered for cost-sharing when the diagnosis is Cystic Fibrosis-Related Diabetes (CFRD) with fasting hyperglycemia. See [Chapter 8, Section 2.3](#) for policy regarding EIPs.

**2.8** Post-operative proton beam radiosurgery/radiotherapy (CPT<sup>2</sup> procedure codes 77520, 77522, 77523, and 77525) may be considered for cost-sharing when the diagnosis is sacral chordoma. See [Chapter 5, Section 3.1](#) for policy regarding proton beam radiosurgery/radiotherapy.

**2.9** Extracorporeal photopheresis (CPT<sup>2</sup> procedure code 36522) may be considered for cost-sharing when the diagnosis is Bronchiolitis Obliterans Syndrome (BOS) that is refractory to immunosuppressive drug treatment. See [Chapter 4, Section 9.2](#) for policy regarding photopheresis.

**2.10** Effective May 1, 2008, the off-label use of Selective Internal Radiation Therapy (SIRT) with yttrium-90 microspheres (resin or glass) may be considered for cost-sharing for the treatment of unresectable liver metastases from neuroendocrine tumors. See [Chapter 5, Section 3.2](#) for policy regarding brachytherapy/radiation therapy.

**2.11** Effective April 15, 2016, Collagen Cross-linking for the treatment of corneal ectasia due to the rare disease Keratoconus is safe and effective and may be considered for cost-sharing.

**2.12** Effective June 1, 2010, Radiofrequency Ablation (RFA), when performed using an U.S. Food and Drug Administration (FDA) approved electro-surgical cutting and coagulation device, may be considered for cost-sharing for the treatment of liver metastases from gastric cancer.

**2.13** Effective September 1, 2012, the NovoTTF-100A system (HCPCS A4555 and E0766) may be cost-shared for the treatment of adult patients (22 years of age or older) with recurrent glioblastoma after surgical and radiation options have been exhausted.

**2.14** Effective February 4, 2011, Radiesse® Voice laryngoplasty injections may be cost-shared for the treatment of type 1 laryngeal cleft (also described as supraglottic interarytenoid defects that extend no further than the true vocal folds).

**2.15** Effective November 27, 1995, Orthotopic Liver Transplantation (OLT) may be cost-shared for the treatment of Crigler-Najjar Syndrome Type I. OLT may be performed both prior to the onset of neurological symptoms or after the onset of neurological symptoms.

**2.16** Effective June 5, 2013, off-label use of intravenous immune globulin for the treatment of Hashimoto's Encephalopathy, may be considered in exceptional circumstances where there is progressive neurologic decline despite appropriate steroid therapy or where steroid therapy is contraindicated.

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**2.17** Effective April 30, 2009, Intrapulmonary Percussive Ventilation (IPV) may be considered for cost-sharing when the diagnosis is Cystic Fibrosis (CF). See [Chapter 8, Section 16.1](#) for policy regarding IPVs.

**2.18** Effective January 4, 2013, allogeneic hematopoietic cell transplant (CPT<sup>3</sup> procedure code 38240) **may be considered for cost-sharing** for the treatment of primary plasma cell leukemia.

**2.19** Effective February 1, 2008, the off-label use of Photodynamic Therapy (CPT<sup>3</sup> procedure code 67221) with Visudyne (HCPCS J3396) may be considered for cost-sharing for the treatment of retinal astrocytic hamartoma in Tuberous Sclerosis.

**2.20** Effective June 25, 2014, intracranial angioplasty with stenting (CPT<sup>3</sup> procedure code 61635) of the venous sinuses may be considered for cost-sharing for the treatment of pseudotumor cerebri (also known as idiopathic intracranial hypertension and benign intracranial hypertension).

**2.21** Effective February 1, 2012, OLT (CPT<sup>3</sup> procedure code 47135) **may be considered for cost-sharing** for the treatment of Acute Intermittent Porphyria.

**2.22** Effective December 1, 2014, Photodynamic Therapy **may be considered for cost-sharing** for the treatment of Central Serous Chorioretinopathy.

**2.23** Effective July 22, 2016, chemotherapy injections (CPT<sup>3</sup> procedure code 96542) may be considered for cost-sharing for the treatment of Central Giant Cell Granuloma (CGCG) of the mandible.

**2.24** Effective July 22, 2016, Peg interferon alfa-2A/180 (HCPCS J3490) may be considered for cost-sharing for the treatment of CGCG of the mandible.

**2.25** Effective August 11, 2016, a Fluorodeoxyglucose (FDG) PET scan (CPT<sup>3</sup> procedure code 78815) may be considered for cost-sharing for the treatment of Takayasu's Arteritis (also known as aortic arch syndrome).

**2.26** Effective August 22, 2016, Gammagard liquid injections (HCPCS J1569) may be considered for cost-sharing for the treatment of branch retinal artery occlusion secondary to Susac's Syndrome.

**2.27** Effective October 13, 2016, an autologous bone marrow transplant, the harvest of autologous stem cells, and the cryopreservation of stem cells may be considered for cost-sharing for the treatment of recurrent medulloblastoma.

**2.28** Effective December 15, 2016, a Magnetic Resonance-guided High Intensity Focused Ultrasound (MRgFUS) may be considered for cost-sharing for the treatment of Desmoid fibromatosis.

**2.29** Effective January 24, 2017, Stereotactic Body Radiation Therapy (SBRT) (CPT<sup>3</sup> procedure codes 77435 and 77373) may be considered for cost-sharing for the treatment of a benign neoplasm of the aortic body and other paraganglia.

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**3.0 EXCLUSIONS**

**3.1** The off-label use of rituximab for the treatment of pediatric linear Immunoglobulin A (IgA) dermatosis is unproven.

**3.2** Proton Beam Therapy (PBT)/radiosurgery/radiotherapy for the treatment of thymoma is unproven.

**3.3** Proton Beam Radiation Therapy (PBRT) for the treatment of juvenile nasal angiofibroma is unproven.

**3.4** TRICARE Overseas Program (TOP) beneficiaries are not subject to the requirements of this policy.

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