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Coverage Policy Number **IP0178**

Repository Corticotropin

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Related Coverage Resources

INSTRUCTIONS FOR USE
The following Coverage Policy applies to health benefit plans administered by Cigna Companies. Certain Cigna Companies and/or lines of business only provide utilization review services to clients and do not make coverage determinations. References to standard benefit plan language and coverage determinations do not apply to those clients. Coverage Policies are intended to provide guidance in interpreting certain standard benefit plans administered by Cigna Companies. Please note, the terms of a customer's particular benefit plan document [Group Service Agreement, Evidence of Coverage, Certificate of Coverage, Summary Plan Description (SPD) or similar plan document] may differ significantly from the standard benefit plans upon which these Coverage Policies are based. For example, a customer's benefit plan document may contain a specific exclusion related to a topic addressed in a Coverage Policy. In the event of a conflict, a customer's benefit plan document always supersedes the information in the Coverage Policies. In the absence of a controlling federal or state coverage mandate, benefits are ultimately determined by the terms of the applicable benefit plan document. Coverage determinations in each specific instance require consideration of 1) the terms of the applicable benefit plan document in effect on the date of service; 2) any applicable laws/regulations; 3) any relevant collateral source materials including Coverage Policies and; 4) the specific facts of the particular situation. Coverage Policies relate exclusively to the administration of health benefit plans. Coverage Policies are not recommendations for treatment and should never be used as treatment guidelines. In certain markets, delegated vendor guidelines may be used to support medical necessity and other coverage determinations..

Overview

This policy supports medical necessity review for repository corticotropin (Acthar Gel®).

Receipt of sample product does not satisfy any criteria requirements for coverage.

Medical Necessity Criteria

Repository corticotropin (Acthar Gel) is considered medically necessary when the following are met:

1. **Treatment of Infantile Spasms.** Individual meets **BOTH** of the following criteria (A and B):
   A. Individual is less than 2 years of age
   B. Medication is prescribed by, or in consultation with, a neurologist

When coverage is available and medically necessary, the dosage, frequency, duration of therapy, and site of care should be reasonable, clinically appropriate, and supported by evidence-based literature and adjusted based upon severity, alternative available treatments, and previous response to therapy.

Reauthorization Criteria

Repository corticotropin (Acthar Gel) is considered medically necessary for continued use when initial criteria are met AND there is documentation of beneficial response.

Authorization Duration

Initial approval duration is up to 1 month.
Reauthorization approval duration is up to 1 month.

Conditions Not Covered

The effectiveness of repository corticotropin (Acthar Gel) has not been demonstrated as clinically superior to conventional corticosteroids and/or immunosuppressive therapy for uses other than infantile spasms, including for the treatment of multiple sclerosis, rheumatic disorders, collagen diseases, dermatologic disorders, allergic states, ophthalmic diseases, respiratory diseases, or edematous states, and is significantly more expensive. Coverage of repository corticotropin may depend on the applicable health benefit plan definition of medical necessity. Where that definition limits coverage to the most cost-effective equivalent treatment, repository corticotropin is not considered medically necessary.

Any other use is considered not medically necessary including the following (this list may not be all inclusive):

1. **Ankylosing Spondylitis.** The American College of Rheumatology guidelines for the treatment of ankylosing spondylitis do not convey a role for ACTH in this condition.[11,12]

2. **Dermatomyositis or Polymyositis.** British Society for Rheumatology guidelines on the management of pediatric, adolescent, and adult individuals with idiopathic inflammatory myopathy (2022) do not cite ACTH as an agent to utilize in individuals with such conditions.[13]

3. **Diabetic Nephropathy.** ACTH is not a cited therapy or the standard of care for the management of chronic kidney disease in individuals with diabetes.[5,14]

4. **Glomerular Kidney Diseases.**
   Examples of diagnoses can include nephrotic syndrome, membranous nephropathy, immunoglobulin A nephropathy, minimal change disease, infection-related glomerulonephritis, focal segmental glomerulosclerosis, and membranoproliferative glomerulonephritis. ACTH is not prominent in related guidelines from KDIGO (2021) and there is a lack of quality evidence regarding ACTH to support its use.[5]

5. **Gout.** American College of Rheumatology guidelines for gout (2020) recommend other therapies beside ACTH for gout flare management (e.g., colchicine, non-steroidal anti-inflammatory drugs, or glucocorticoids).[9]

6. **Juvenile Idiopathic Arthritis.** Related guidelines from the American College of Rheumatology regarding the treatment of juvenile idiopathic arthritis (2021) do not mention ACTH as having a role for this disease.[15]

7. **Lupus Nephritis.** The KDIGO guidelines for the management of glomerular disease (2021) cite many other agents besides ACTH for the management of this condition.[5] The European League Against Rheumatism-European Renal Association-European Dialysis and Transplantation Association joint

recommendations on the management of lupus nephritis do not cite ACTH as a therapy to use in this condition.[16]

8. **Multiple Sclerosis, Acute Exacerbations.** High-dose corticosteroids, usually short-term intravenous methylprednisolone, are the accepted standard of care short-term for acute relapses or exacerbations.[6]

9. **Ophthalmic Conditions.** Only limited data describe the use of ACTH in ophthalmic-related conditions (e.g., acute optic neuritis, keratitis, retinal vasculitis).[2,17-19] Prospective data are needed to more rigorously define the efficacy and safety of ACTH in ocular disease.

10. **Psoriatic Arthritis.** The American College of Rheumatology/National Psoriasis Foundation guidelines for the treatment of psoriatic arthritis (2018) do not mention a role for ACTH in this condition.[20]

11. **Rheumatoid Arthritis.** The American College of Rheumatology guidelines for the treatment of rheumatoid arthritis (2021) do not mention a role for ACTH in this disease state.[21]

12. **Sarcoidosis.** The European Respiratory Society published guidelines on the treatment of sarcoidosis (2021).[10] Repository corticotropin use should be reserved for individuals who have failed prior treatments (e.g., steroids, antimetabolites). Only limited data are available. Repository corticotropin should be considered in a case by case basis only when other therapies are not effective or tolerated.

Coding / Billing Information

Note: 1) This list of codes may not be all-inclusive.
2) Deleted codes and codes which are not effective at the time the service is rendered may not be eligible for reimbursement.

Considered Medically Necessary when criteria in the applicable policy statements listed above are met:

HCPCS Codes	Description
J0800	Injection, corticotropin, up to 40 units

Background

OVERVIEW
Acthar, an adrenocorticotropic hormone (ACTH) analog, is indicated for the following uses:[1]

- **Infantile spasms**, treatment of, in infants and children < 2 years of age.
- **Multiple sclerosis, treatment of exacerbations** in adults.

Although data are limited, the prescribing information notes that Acthar may also be used for the following disorders and diseases:[1]

- **Allergic states**, such as serum sickness.
- **Collagen diseases**, during an exacerbation or as a maintenance therapy in selected cases of systemic lupus erythematosus and systemic dermatomyositis (polymyositis).
- **Dermatologic diseases**, such as severe erythema multiforme and Stevens-Johnson syndrome.
- **Edematous state** including to induce a diuresis or a remission of proteinuria in the nephrotic syndrome without uremia of the idiopathic type or that due to lupus erythematosus.
- **Respiratory diseases** such as symptomatic sarcoidosis.
- **Rheumatoid disorders**, as an adjunctive therapy for short-term administration (to tide the patient over an acute episode or exacerbation) in psoriatic arthritis, rheumatoid arthritis (including juvenile rheumatoid arthritis) [selected cases may require low-dose maintenance therapy], and ankylosing spondylitis.

- **Ophthalmic diseases** including severe acute and chronic allergic and inflammatory processes involving the eye and its adnexa such as keratitis, iritis, iridocyclitis, diffuse posterior uveitis and choroiditis, optic neuritis, chorioretinitis, and anterior segment inflammation.

Clinical Efficacy
A review regarding repository corticotropin found few randomized controlled trials supporting the clinical benefit of repository corticotropin or ACTH for various conditions (e.g., use in rheumatoid arthritis, ankylosing spondylitis, optic neuritis, systemic lupus erythematosus, and nephrotic syndrome).[2] Most data suggest that repository corticotropin or ACTH was not superior to corticosteroids for treating relapses in patients with multiple sclerosis.

Guidelines
Several guidelines discuss repository corticotropin or ACTH.

- The **American Academy of Neurology and the Child Neurology Society** published an evidence-based guideline for the medical treatment of infantile spasms (2012).[3] ACTH is a first-line agent for the short-term treatment of infantile spasms.
- **Infantile Spasms Working Group** published a US consensus report on infantile spasms in 2010.[4] Most patients with this condition (90%) present within the first year of life. ACTH is an effective first-line therapy for infantile spasms.
- **Kidney Disease Improving Global Outcomes (KDIGO) published clinical practice guidelines for the management of glomerular disease (2021**).[5] This includes diagnoses such as nephrotic syndrome, membranous nephropathy, immunoglobulin A nephropathy, minimal change disease, infection-related glomerulonephritis, focal segmental glomerulosclerosis, membranoproliferative glomerulonephritis, and lupus nephritis. ACTH is not prominent in the guidelines and there is a lack of quality evidence regarding ACTH.
- **National Multiple Sclerosis Society** has recommendations regarding corticosteroids in the management of multiple sclerosis relapses or exacerbations.[6] High-dose corticosteroids are the accepted standard of care short-term. The most common regimen is 500 to 1,000 mg of intravenous methylprednisolone given daily for 3 to 5 days, with or without an oral steroid tapering regimen (most often prednisone) for 1 to 3 weeks. ACTH and high-dose intravenous methylprednisolone have been shown to possess similar efficacy in the management of multiple sclerosis relapses.[7]
- The **American College of Rheumatology** has many guidelines regarding use in rheumatoid-type conditions.[8] ACTH does not have a prominent role and is generally not recommended for use in any of the related American College of Rheumatology guidelines.
- **The American College of Rheumatology has guidelines for the management of gout** (2020).[9] For gout flare management, using colchicine, non-steroidal anti-inflammatory drugs, or glucocorticoids (oral, intraarticular, or intramuscular) are appropriate first-line therapy for gout flare over interleukin-1 inhibitors or ACTH.
- **The European Respiratory Society published guidelines on the treatment of sarcoidosis (2021)**.[10] Repository corticotropin use should be reserved for patients who have failed prior treatments (e.g., steroids, antimetabolites). Only limited data are available. Repository corticotropin should be considered in a case by case basis only when other therapies are not effective or tolerated.

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