

CODING AND REIMBURSEMENT GUIDE

ZEMAIRA[®] (Alpha₁-Proteinase Inhibitor [Human])

CODING INFORMATION: ZEMAIRA® (ALPHA₁-PROTEINASE INHIBITOR [HUMAN])

Disclaimer: The codes being provided are from a complex and evolving medical coding system. The treating physician is solely responsible for diagnosis coding and determination of the appropriate ICD-10-CM codes that describe the patient's condition and are supported by the medical record. All codes provided are for informational purposes and are not an exhaustive list. The CPT, HCPCS, ICD-10-CM, and ICD-10-PCS codes provided are based on AMA or CMS guidelines. The billing party is solely responsible for coding of services (eg, CPT Coding). Because government and other third-party payer coding requirements change periodically, please verify current coding requirements directly with the payer being billed.

ICD-10-CM Diagnosis Code for Alpha-1 Antitrypsin Deficiency¹

ICD-10-CM Code	Description
E88.01	Alpha-1-antitrypsin (AAT) deficiency
Some payers will only cover treatment for Alpha-1 antitrypsin deficiency when the patient also has documented panlobular emphysema. Code only if applicable:	
J43.1	Panlobular emphysema [panacinar emphysema]

HCPCS Codes²:

The following drug code has been assigned by the Centers for Medicare & Medicaid Services National Level II HCPCS coding system and can be used for ZEMAIRA. Providers are advised to verify correct code(s) with the payer.

HCPCS Code	Description
J0256	Injection, alpha 1 proteinase inhibitor (human), not otherwise specified, 10 mg

ZEMAIRA is supplied in a single-dose vial containing approximately 1000 mg, 4000 mg, or 5000 mg of functionally active A₁-PI as a white to off-white lyophilized powder for reconstitution with Sterile Water for Injection, USP. The amount of functional A₁-PI is printed on the vial label and carton.

Note: When using the HCPCS code, include the drug NDC for ZEMAIRA, and the total number of units printed on the carton. If the number of units is not a multiple of the 10 mg units provided in the long HCPCS descriptor for J0256, provider shall round up to the nearest whole number to express the number as a multiple.³

JW and JZ Modifier³:

For Medicare, providers are required to use the JW modifier for claims with unused drugs or biologicals from single use vials that are appropriately discarded. The amount discarded must be documented in the patient's medical record.

Effective July 1, 2023, Medicare requires the JZ modifier on all claims for single-dose containers where there are no discarded amounts. Providers are advised to verify requirements with non-Medicare payers.

"Special Payment Instructions" for J0256⁴:

Status indicators (SI) describe how particular HCPCS codes and APCs are paid under the Outpatient Prospective Payment System (OPPS). J0256 for ZEMAIRA has **Status Indicator K**, which is defined as non-pass-through drugs/implantable biologicals including therapeutic radiopharmaceuticals.

As a result, when administered in an outpatient facility, payment for ZEMAIRA is separate from payment for administration under the OPPS, because it exceeds the per day cost threshold for separately payable drugs. These payment rates are published quarterly on the Centers for Medicare and Medicaid Services (CMS) website at <https://www.cms.gov/Medicare/Coding/HCPCSReleaseCodeSets/HCPCS-Quarterly-Update>.

ZEMAIRA National Drug Code (NDC)

Certain payers may require separate prior authorizations if patient's dose requires use of multiple kit sizes. For example, a patient receiving a 6000 mg dose may need both a 5000 mg and a 1000 mg vial, which could require separate prior authorizations. Payers may also require NDC #s on claims, which will require billing each kit size on separate lines of the claim form. Providers are advised to verify requirements with the payer.

ZEMAIRA Kit NDC #	Presentation	Billing Format
0053-7201-02	1000 mg of functionally active A ₁ -PI	00053-7201-02
0053-7202-02	4000 mg of functionally active A ₁ -PI	00053-7202-02
0053-7203-02	5000 mg of functionally active A ₁ -PI	00053-7203-02

RED "0" is required for some claims file formats (11-digit, 5-4-2 formatting). Use if applicable.

CPT Administration Codes

If the provider performs administration services in conjunction with a ZEMAIRA infusion, the following administration codes may be billed for this service. Providers are advised to verify correct code(s) with the payer:

Please verify current coding requirements directly with the payer being billed.

CPT Code	Description
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour ⁵
OR	
96374	Intravenous push, single or initial substance/drug ⁶
NOTE: An IV push is defined as: (a) injection in which the individual who administers the drug/substance is continuously present to administer the injection and observe the patient, or (b) an infusion of 15 minutes or less.	
NOTE: The recommended dosage of 60 mg/kg body weight will take approximately 15 minutes to infuse (based on an "average" 75 kg, 165 lb patient). Individual experiences may vary.	
S9346	Home infusion therapy, alpha-1 proteinase inhibitor (eg, prolastin); administrative services, professional pharmacy services, care coordination, and all necessary supplies and equipment (drugs and nursing visits coded separately), per diem ⁷
Note from HCPCS Level II Manual: Noncovered by Medicare	
Note from CPT National Correct Coding Initiative (NCCI) Policy Manual for Medicare Services: Drug administration CPT code 96365 and 96366 have been valued to include the work and practice expenses of CPT code 99211. Therefore, CPT code 99211 is not reportable with CPT code 96372. Other non-facility based evaluation and management codes may be separately reportable with modifier 25 if the physician provides a significant and separately identifiable E&M service.	


Payers may allow administration of ZEMAIRA by HCPs in facility outpatient settings, as well as in the home. The benefit category for coverage (medical or pharmacy), may differ depending on site of care, and should be checked before claim submission.

For Medicare beneficiaries, administration of intravenous (IV) drugs requires the supervision of a healthcare professional (HCP). Both ZEMAIRA and its administration are covered as a Part B (Medical) service when performed in a facility-based outpatient setting. Note that payers other than Medicare may require patients to meet additional medical necessity criteria for approval of facility-based services.

For Medicare beneficiaries, when ZEMAIRA is given at home, the drug may be covered under either Part B or Part D, depending on circumstances.

For reimbursement support, please call ZEMAIRA Connect at 1-866-ZEMAIRA or 1-866-936-2472.

CLAIM FORM EXAMPLE



CARRIER

HEALTH INSURANCE CLAIM FORM

APPROVED BY NATIONAL UNIFORM CLAIM COMMITTEE (NUCC) 02/12

<input type="checkbox"/> <input type="checkbox"/> PICA										<input type="checkbox"/> <input type="checkbox"/> PICA																																							
1. MEDICARE <input type="checkbox"/> (Medicare#)					MEDICAID <input type="checkbox"/> (Medicaid#)					TRICARE <input type="checkbox"/> (ID#/DoD#)					CHAMPVA <input type="checkbox"/> (Member ID#)					GROUP HEALTH PLAN <input type="checkbox"/> (ID#)					FECA BLK LUNG <input type="checkbox"/> (ID#)					OTHER <input type="checkbox"/> (ID#)					1a. INSURED'S LD. NUMBER (For Program in Item 1)														
2. PATIENT'S NAME (Last Name, First Name, Middle Initial)															3. PATIENT'S BIRTH DATE MM DD YY					SEX M <input type="checkbox"/> F <input type="checkbox"/>					4. INSURED'S NAME (Last Name, First Name, Middle Initial)																								
5. PATIENT'S ADDRESS (No., Street)															6. PATIENT RELATIONSHIP TO INSURED Self <input type="checkbox"/> Spouse <input type="checkbox"/> Child <input type="checkbox"/> Other <input type="checkbox"/>										7. INSURED'S ADDRESS (No., Street)																								
CITY					STATE					8. RESERVED FOR NUCC USE										CITY					STATE																								
ZIP CODE					TELEPHONE (Include Area Code)															ZIP CODE					TELEPHONE (Include Area Code)																								
9. OTHER INSURED'S NAME (Last Name, First Name, Middle Initial)															10. IS PATIENT'S CONDITION RELATED TO:										11. INSURED'S POLICY GROUP OR FECA NUMBER																								
a. OTHER INSURED'S POLICY OR GROUP NUMBER					a. EMPLOYMENT? (Current or Previous)					a. INSURED'S DATE OF BIRTH					SEX																																		
b. RESERVED FOR NUCC USE					<input type="checkbox"/> YES <input type="checkbox"/> NO					b. AUTO ACCIDENT?					PLACE (State)					b. OTHER CLAIM ID (Designated by NUCC)																													
c. RESERVED FOR NUCC USE					<input type="checkbox"/> YES <input type="checkbox"/> NO					c. OTHER ACCIDENT?					<input type="checkbox"/> YES <input type="checkbox"/> NO					c. INSURANCE PLAN NAME OR PROGRAM NAME																													
d. INSURANCE PLAN NAME OR PROGRAM NAME															10d. CLAIM CODES (Designated by NUCC)										d. IS THERE ANOTHER HEALTH BENEFIT PLAN?																								
																									<input type="checkbox"/> YES <input type="checkbox"/> NO <i>If yes, complete items 9, 9a, and 9d.</i>																								
READ BACK OF FORM BEFORE COMPLETING & SIGNING THIS FORM.															12. PATIENT'S OR AUTHORIZED PERSON'S SIGNATURE I authorize the release of any medical or other information necessary to process this claim. I also request payment of government benefits either to myself or to the party who accepts assignment below.										13. INSURED'S OR AUTHORIZED PERSON'S SIGNATURE I authorize payment of medical benefits to the undersigned physician or supplier for services described below.																								
SIGNED _____															DATE _____										SIGNED _____																								
14. DATE OF CURRENT ILLNESS, INJURY, or PREGNANCY (LMP)					15. OTHER DATE					16. DATES PATIENT UNABLE TO WORK IN CURRENT OCCUPATION																																							
MM DD YY					MM DD YY					FROM MM DD YY TO MM DD YY																																							
17. NAME OF REFERRING PROVIDER OR OTHER SOURCE															17a. _____					17b. NPI					18. HOSPITALIZATION DATES RELATED TO CURRENT SERVICES																								
																									FROM MM DD YY TO MM DD YY																								
19. ADDITIONAL CLAIM INFORMATION (Designated by NUCC)															20. OUTSIDE LAB? \$ CHARGES																																		
															<input type="checkbox"/> YES <input type="checkbox"/> NO																																		
21. DIAGNOSIS OR NATURE OF ILLNESS OR INJURY Relate A-L to service line below (24E)															22. RESUBMISSION CODE										ORIGINAL REF. NO.																								
A. _____ B. _____ C. _____ D. _____																																																	
E. _____ F. _____ G. _____ H. _____																																																	
I. _____ J. _____ K. _____ L. _____																																																	
24A. DATE(S) OF SERVICE					B. PLACE OF SERVICE					C. EMG					D. PROCEDURES, SERVICES, OR SUPPLIES (Explain Unusual Circumstances)					E. DIAGNOSIS POINTER					F. \$ CHARGES					G. DAYS OR UNITS					H. EFSBT Family Plan					I. ID. QUAL.					J. RENDERING PROVIDER ID. #				
From MM DD YY To MM DD YY																																																	
1															24D					24E					24G					NPI																			
2																														NPI																			
3																														NPI																			
4																														NPI																			
5																														NPI																			
6																														NPI																			
25. FEDERAL TAX ID. NUMBER					SSN EIN					26. PATIENT'S ACCOUNT NO.					27. ACCEPT ASSIGNMENT? (For govt. claims, see back)					28. TOTAL CHARGE \$					29. AMOUNT PAID \$					30. Rsvd for NUCC Use																			
31. SIGNATURE OF PHYSICIAN OR SUPPLIER INCLUDING DEGREES OR CREDENTIALS (I certify that the statements on the reverse apply to this bill and are made a part thereof.)															32. SERVICE FACILITY LOCATION INFORMATION										33. BILLING PROVIDER INFO & PH # ()																								
SIGNED _____															DATE _____										a. NPI					b. _____					a. NPI					b. _____									

NUCC Instruction Manual available at: www.nucc.org PLEASE PRINT OR TYPE APPROVED OMB-0938-1197 FORM 1500 (02-12)

CLAIM FORM EXAMPLE (CONTINUED)

Field 19: Additional Claim Information

HCPCS code (J0256) can be used for other alpha-1 proteinase inhibitors besides ZEMAIRA. Therefore, when billing ZEMAIRA, the provider must include additional information about this specific product in a free-text field (eg, Field 19 or its electronic equivalent).

Payers typically require: drug name, NDC, route of administration, and amount used for treatment. For example:

ZEMAIRA NDC 00053-7201-02; Intravenous, 1000 mg of functionally active A₁-PI

ZEMAIRA NDC 00053-7202-02; Intravenous, 4000 mg of functionally active A₁-PI

ZEMAIRA NDC 00053-7203-02; Intravenous, 5000 mg of functionally active A₁-PI

NDC Code: Note that the 10-digit NDC code for ZEMAIRA is converted to an 11-digit billing format by inserting a zero in the first segment. The NDC code on the package is XXXX-XXXX-XX; the 11-digit billing format is 0XXXX-XXXX-XX.

Field 21: Diagnosis or Nature of Illness of Injury

Enter the ICD-10-CM diagnosis code(s) that best describes the reason the patient is receiving care.

Field 24A (Shaded Area): NDC

For Medicaid claims, and for Medicare claims that will cross over to Medicaid as the secondary payer, the NDC in a specific format is required in the shaded area above the line on which ZEMAIRA is reported in Field 24A. **Various state Medicaid plans and Medicare may have different reporting formats for this information.**

Usually, the provider will need to supply the NDC in 11-digit format preceded by the modifier N4 (N40XXXX-XXXX-XX). This is typically followed by the NDC unit of measure (F2 [mg]) and the numeric quantity of the NDC that was dispensed.

Other payers may require different information. Check with the payer for specific requirements.

Field 24D: CPT/HCPCS

HCPCS coding for ZEMAIRA should use:

J0256: Injection, alpha 1 proteinase inhibitor (human), not otherwise specified, 10 mg

For Medicare beneficiaries, the following code may not be payable.

- S9346—Home infusion therapy, alpha-1 proteinase inhibitor (eg, prolastin); administrative services, professional pharmacy services, care coordination, and all necessary supplies and equipment (drugs and nursing visits coded separately), per diem

The code selected is based on the documented service provided to the patient.

Field 24E: Diagnosis Pointer

Enter the number(s) from Field 21 that best describes the medical necessity for the service.

Field 24G: Days or Units

J0256: Injection, alpha 1 proteinase inhibitor (human), not otherwise specified, 10 mg

Requirements for Field 24G vary by payer. For example, some payers rely on the information in Field 19 for this data and request a placeholder of "1" in Field 24G. Other payers may require 1 unit per unit of drug used in the care of the patient.

INDICATIONS AND IMPORTANT SAFETY INFORMATION

ZEMAIRA®, Alpha₁-Proteinase Inhibitor (Human), is indicated for chronic augmentation and maintenance therapy for adults with alpha₁-proteinase inhibitor (A₁-PI) deficiency and emphysema. The effect of augmentation therapy with ZEMAIRA or any A₁-PI product on pulmonary exacerbations and progression of emphysema in A₁-PI deficiency has not been demonstrated in randomized, controlled clinical studies.

ZEMAIRA is not indicated for lung disease patients in whom severe A₁-PI deficiency has not been established.

ZEMAIRA is contraindicated in patients with a history of severe systemic reactions to the product or to A₁-PI protein, including anaphylaxis. Due to the risk of severe hypersensitivity, ZEMAIRA is also contraindicated in immunoglobulin A-deficient patients with antibodies against IgA.

Use caution in administering ZEMAIRA to patients who have experienced anaphylaxis or severe systemic reactions to another A₁-PI product. Patients with selective or severe IgA deficiency can develop antibodies to IgA and are at greater risk of such reactions. If anaphylactic or severe anaphylactoid reactions occur during infusion, discontinue immediately.

In pre-licensure clinical studies, the following adverse reactions were reported in at least 5% of subjects receiving ZEMAIRA: headache, sinusitis, upper respiratory infection, bronchitis, asthenia, increased cough, fever, injection-site hemorrhage, rhinitis, sore throat, and vasodilation.

ZEMAIRA is derived from human plasma. The risk of transmission of infectious agents, including viruses and, theoretically, the Creutzfeldt-Jakob disease (CJD) agent and its variant (vCJD), cannot be completely eliminated.

Please see full prescribing information for ZEMAIRA on page 7.

To report SUSPECTED ADVERSE REACTIONS, contact the CSL Behring Pharmacovigilance Department at 1-866-915-6958 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch.

References: **1.** National Center for Health Statistics (US Department of Health and Human Services) International Classification of Diseases, Tenth Revision, Clinical Modification (ICD-10-CM). Comprehensive Listing ICD-10-CM Files: 2024 release of ICD-10-CM. <https://www.cdc.gov/nchs/icd/comprehensive-listing-of-icd-10-cm-files.htm>. Accessed September 15, 2023. **2.** U.S. Centers for Medicare & Medicaid Services (US Department of Health and Human Services). HCPCS Quarterly Update. <https://www.cms.gov/medicare/coding-billing/healthcare-common-procedure-system/quarterly-update>. Accessed September 15, 2023. **3.** Billing and Coding: JW and JZ Modifier Billing Guidelines. Article A55932. <https://www.cms.gov/medicare-coverage-database/view/article.aspx?articleid=55932>. Accessed September 15, 2023. **4.** MLN Matters MM10923, October 2018 Update of the Hospital Outpatient Prospective Payment System (OPPS). US Department of Health and Human Services, Centers for Medicare and Medicaid Services, Medicare Learning Network. <https://www.cms.gov/Outreach-and-Education/Medicare-Learning-Network-MLN/MLNMattersArticles/downloads/MM10923.pdf>. Accessed September 15, 2023. **5.** 2023 CPT Code 96365. American Academy of Professional Coders website. <https://www.aapc.com/codes/cpt-codes/96365>. Accessed September 15, 2023. **6.** 2023 CPT Code 96374. American Academy of Professional Coders website. <https://www.aapc.com/codes/cpt-codes/96374>. Accessed September 15, 2023. **7.** HCPCS Codes. Temporary National Codes (Non-Medicare). Codify by AAPC. <https://www.aapc.com/codes/hcpcs-codes/S9346>. Accessed September 15, 2023.

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1020 First Avenue, PO Box 61501, King of Prussia, PA 19406-0901 USA
www.CSLBehring.com www.ZEMAIRA.com USA-ZMR-0042-OCT23

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