Kanuma®
(sebelipase alfa)
intravenous infusion
20mg/
10mL

Coding and Billing Guide for the Use of KANUMA

INDICATION

KANUMA® (sebelipase alfa) is indicated for the treatment of patients with a diagnosis of Lysosomal Acid Lipase (LAL) deficiency.

IMPORTANT SAFETY INFORMATION INCLUDING BOXED WARNING

WARNING: HYPERSENSITIVITY REACTIONS INCLUDING ANAPHYLAXIS

Patients treated with enzyme replacement therapies have experienced life-threatening hypersensitivity reactions, including anaphylaxis. Anaphylaxis has occurred during the early course of enzyme replacement therapy and after extended duration of therapy.

Initiate KANUMA in a healthcare setting with appropriate medical monitoring and support measures, including access to cardiopulmonary resuscitation equipment. If a severe hypersensitivity reaction (e.g., anaphylaxis) occurs, discontinue KANUMA and immediately initiate appropriate medical treatment, including use of epinephrine. Inform patients of the symptoms of life-threatening hypersensitivity reactions, including anaphylaxis and to seek immediate medical care should symptoms occur [see Warnings and Precautions (5.1)].

Please see Important Safety Information on pages 1, 9, and 10 and full Prescribing Information for KANUMA® (sebelipase alfa), including Boxed WARNING.

US/KAN-L/0042 V3 05/2024

PRODUCT OVERVIEW[1]

KANUMA® (sebelipase alfa) is indicated for the treatment of patients with a diagnosis of Lysosomal Acid Lipase (LAL) deficiency.

KANUMA is a concentrated solution administered as an intravenous (IV) infusion after dilution over at least 2 hours in most patients.

KANUMA is supplied as 20 mg/10 mL (2 mg/mL) solution in single-dose vials.

Infusions for KANUMA should be administered by a healthcare professional in the hospital inpatient setting, hospital outpatient clinic, or physician office.

PURPOSE OF THIS GUIDE

Alexion Pharmaceuticals, Inc. has developed the KANUMA Coding and Billing Guide to provide objective and publicly available coding and billing information.

This document is provided for informational purposes only and is not legal advice or official guidance from payers. It is not intended to increase or maximize reimbursement. Alexion does not warrant, promise, guarantee, or make any statement that the use of this information will result in coverage or payment for KANUMA, or that any payment received will cover providers' costs. Alexion is not responsible for any action providers take in billing for, or appealing, KANUMA claims.

Hospitals and physicians are responsible for compliance with Medicare and other payer rules and requirements and for the information submitted with all claims and appeals. Before any claims or appeals are submitted, hospitals and physicians should review official payer instructions and requirements, should confirm the accuracy of their coding or billing practices with these payers, and should use independent judgment when selecting codes that most appropriately describe the services or supplies provided to a patient.

Please visit **kanuma.com/hcp** for additional information or for inquiries regarding reimbursement, please call 1-888-765-4747 to speak with a OneSource™ patient support specialist who can connect you with your local Field Reimbursement Manager (FRM). OneSource™ is available Monday through Friday, 8:30 AM - 8 PM EST.

Please see Important Safety Information on pages 1, 9, and 10 and full Prescribing Information for KANUMA® (sebelipase alfa), including Boxed WARNING.

CODING FOR KANUMA® (sebelipase alfa) IN LYSOSOMAL ACID LIPASE DEFICIENCY (LAL-D)

Diagnosis Coding

The following International Classification of Diseases, 10th Revision, Clinical Modification (ICD-10-CM) code may be appropriate to describe patients diagnosed with LAL-D:

ICD-10-CM Diagnosis Code[2]	Code Descriptor
E75.5	Other lipid storage disorders

Drug Coding

The following drug-specific Healthcare Common Procedure Coding System (HCPCS) billing code can be reported on medical claim forms to payers:

HCPCS Code[3]	Code Descriptor
J2840	Injection, sebelipase alfa, 1 mg

The Centers for Medicare & Medicaid Services (CMS) is requiring the uniform use of the JW (Drug Amount Discarded/Not Administered to Any Patient) modifier to report discarded drugs and biologicals under Medicare Part B. When submitting claims for unused drugs or biologicals from single-use vials that are appropriately discarded, the provider must report the unused portion on a separate claim line from the amount administered to the patient and apply the JW modifier to the HCPCS code for the unused portion. The discarded amounts of drug or biological should be documented in the patient's medical record.[4]

Some payers, including Medicaid, require drugs like KANUMA to be billed on medical claims with the product's National Drug Code (NDC) in addition to the HCPCS code. Payers typically require healthcare professionals to use the Health Insurance Portability and Accountability Act (HIPAA)-compliant, 11-digit NDC format[5]:

11-Digit NDC[1,5]	Code Descriptor	Strength
25682-0007-01	KANUMA® (sebelipase alfa) single-use vial	20 mg/10 mL

Please note that payers have different guidance for placement of the NDC on medical claims. Typically, the 11-digit NDC is reported without any dashes or other punctuation.[5]

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CODING FOR KANUMA® (sebelipase alfa) IN LAL-D

Drug Administration Services

Payers may offer separate coverage and reimbursement for drug administration services. The following are possible International Classification of Diseases, 10th Revision, Procedure Coding System (ICD-10-PCS) procedure codes to report the administration of KANUMA in the hospital inpatient setting:

ICD-10-PCS Code[6]	Code Descriptor
3E033GC	Introduction of other therapeutic substance into peripheral vein, percutaneous approach
3E043GC	Introduction of other therapeutic substance into central vein, percutaneous approach

The following Current Procedural Terminology (CPT®) codes may be appropriate to report administration of KANUMA in physician offices and hospital outpatient clinics. Individual payer policies should be reviewed for reporting requirements.

CPT Code[7]	Code Descriptor
96365	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); initial, up to 1 hour
96366	Intravenous infusion, for therapy, prophylaxis, or diagnosis (specify substance or drug); each additional hour (list separately in addition to code for primary procedure)

KANUMA in the patient home[a]:

CPT Code[7]	Code Descriptor
99601	Home infusion/specialty drug administration, per visit (up to 2 hours)
99602	Home infusion/specialty drug administration, per visit (up to 2 hours); each additional hour (list separately in addition to code for primary procedure)

[a]This is not an all-inclusive list; codes for home services may vary by payer. Check individual payer requirements for reporting.

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CLAIM FORMS

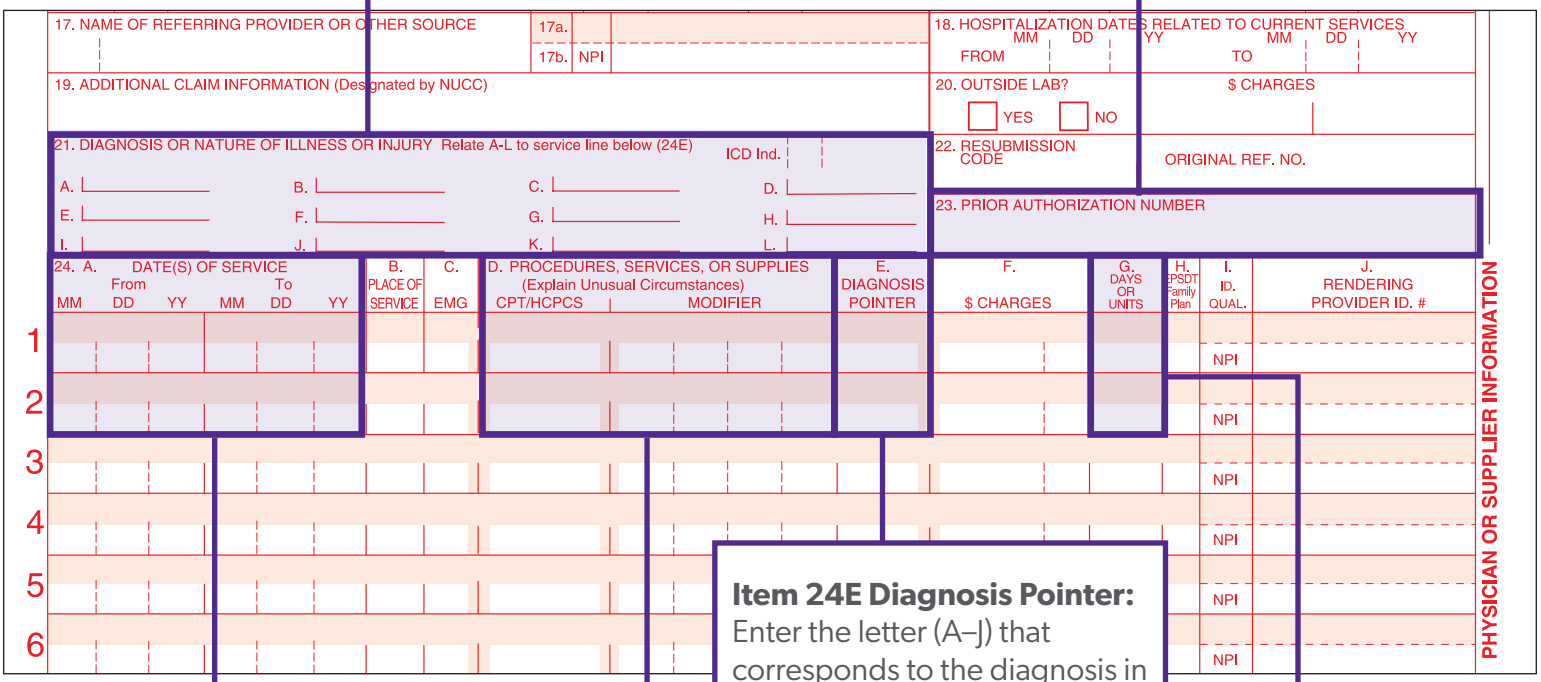
Sample CMS-1500 (or the electronic equivalent 837P): Physician Office[8]

For an example of a completed CMS-1500 form, go to page 6.

Item 21 Diagnosis: Enter the appropriate diagnosis code, eg,

- ICD-10-CM **E75.5** Other lipid storage disorder

Item 23 Prior Authorization: Enter the prior authorization number assigned by the payer.



Item 24E Diagnosis Pointer: Enter the letter (A–J) that corresponds to the diagnosis in Item 21.

Item 24A Date(s) of Service: Enter the NDC number in the shaded area and the month, day, and year in the white space below.

Note: Check payer requirements and format for reporting NDC.

Item 24D Procedures/Services/Supplies: Enter the appropriate CPT/HCPCS codes and modifiers, eg,

- Drug: **J2840** for KANUMA® (sebelipase alfa) per 1 mg
- Administration: **96xxx** for drug administration

Note: Some payers may provide specific guidance.

Item 24G Units: Enter the appropriate number of units of service, eg, KANUMA 200 mg is reported with "200" billing units.

Please see Important Safety Information on pages 1, 9, and 10 and full Prescribing Information for KANUMA® (sebelipase alfa), including Boxed WARNING.

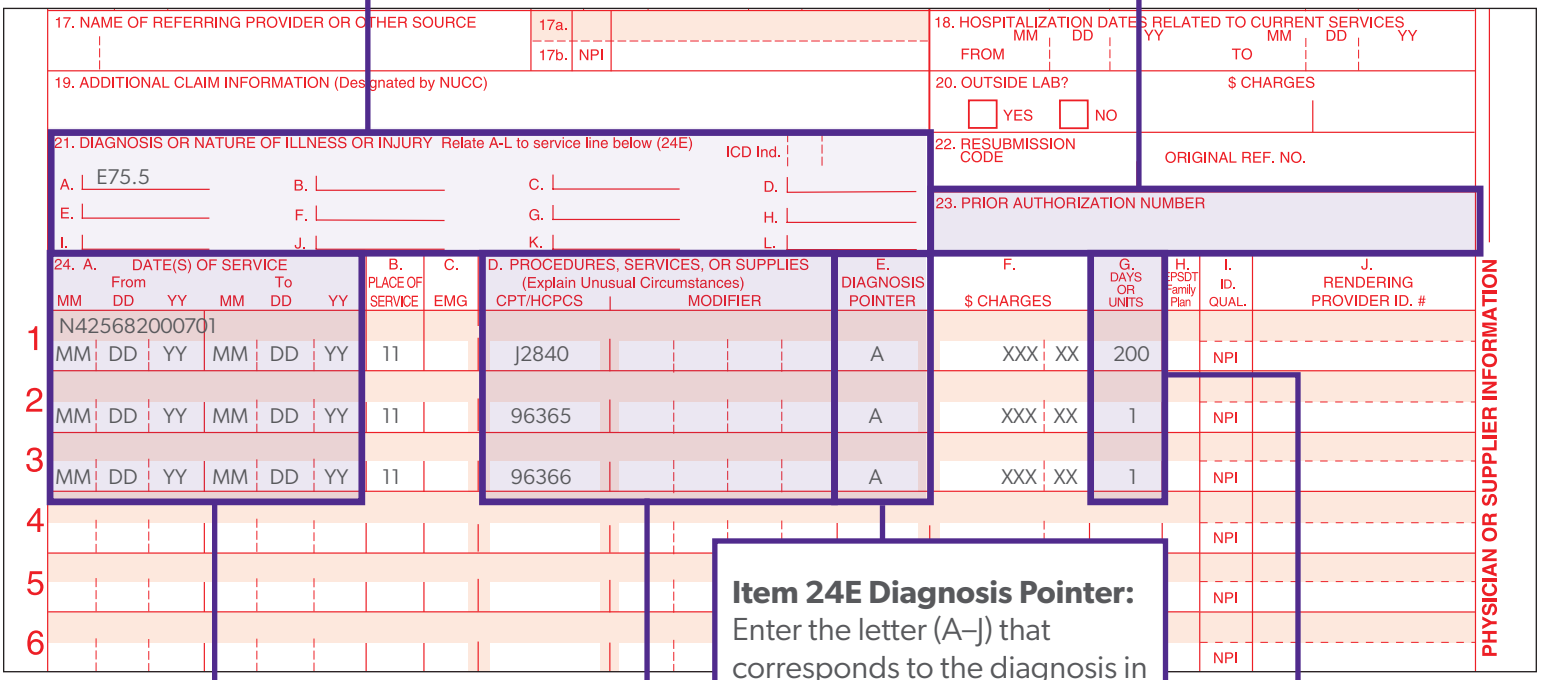
Sample CMS-1500 (or the electronic equivalent 837P): Physician Office[8]

Example claim form for a 200 mg KANUMA® (sebelipase alfa) IV infusion:

Item 21 Diagnosis: Enter the appropriate diagnosis code, eg,

- ICD-10-CM **E75.5** Other lipid storage disorder

Item 23 Prior Authorization: Enter the prior authorization number assigned by the payer.



Item 24E Diagnosis Pointer: Enter the letter (A–J) that corresponds to the diagnosis in Item 21.

Item 24A Date(s) of Service: Enter the NDC number in the shaded area and the month, day, and year in the white space below. The "N4" qualifier is required before the NDC; do not include dashes.

Item 24D Procedures/Services/Supplies: Enter the appropriate CPT/HCPCS codes and modifiers, eg,

- Drug: **J2840** for KANUMA® (sebelipase alfa) per 1 mg
- Administration: **96xxx** for drug administration

Note: Some payers may provide specific guidance.

Item 24G Units: Enter the appropriate number of units of service, eg, KANUMA 200 mg is reported with "200" billing units.

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Sample CMS-1450 (UB-04) (or the electronic equivalent 837I): Hospital Outpatient Clinic or Facility[9]

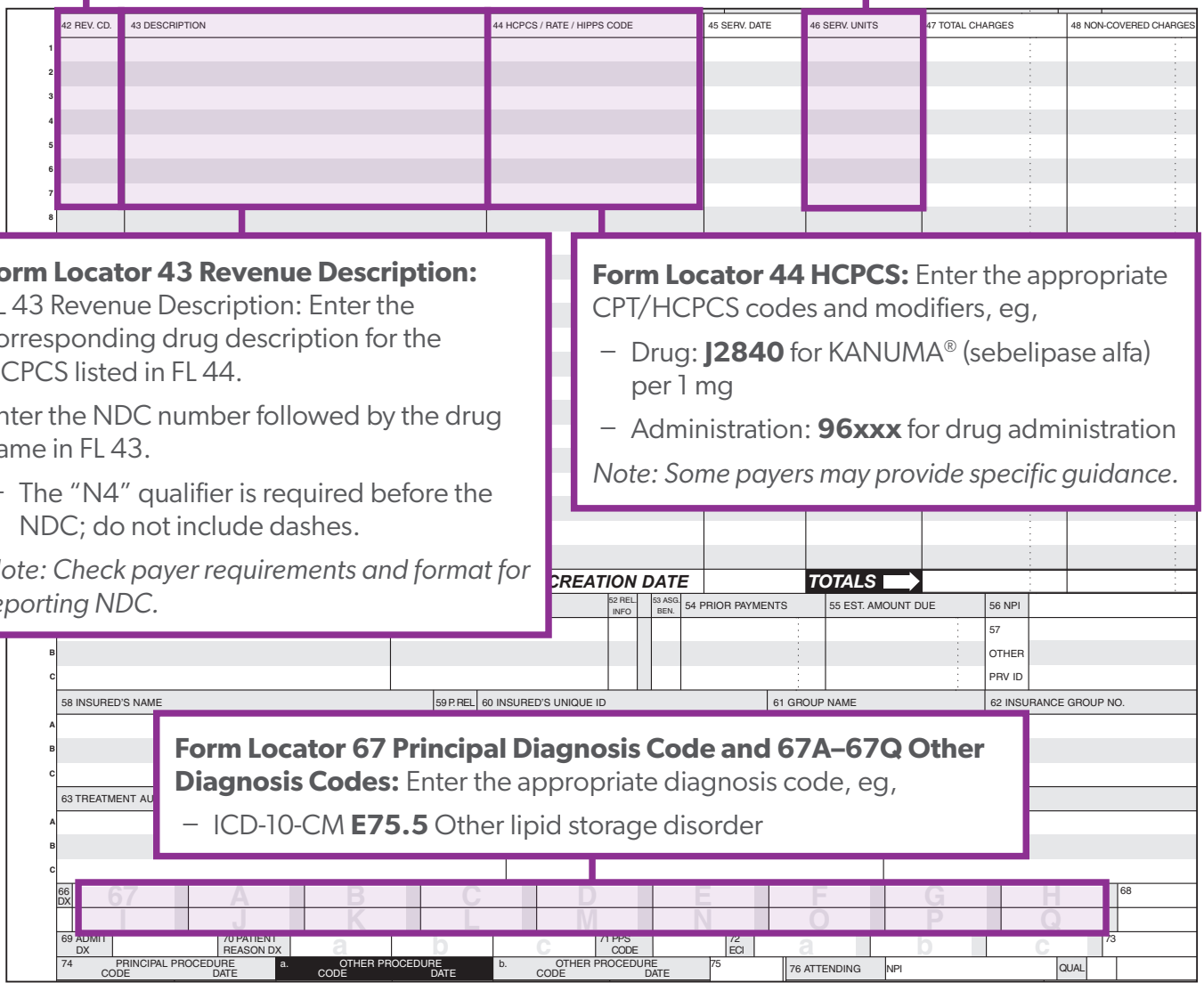
For an example of a completed CMS-1450 form, go to page 8.

Form Locator 42 Revenue Code: Enter the appropriate revenue code,[10] eg,

- **0636** for KANUMA® (sebelipase alfa)
- **0260** for IV infusion

Note: Other revenue codes may apply.

Form Locator 46 Units of Service: Enter the appropriate number of units of service, eg, KANUMA 200 mg is reported with "200" billing units.



Form Locator 43 Revenue Description: FL 43 Revenue Description: Enter the corresponding drug description for the HCPCS listed in FL 44.

Enter the NDC number followed by the drug name in FL 43.

- The "N4" qualifier is required before the NDC; do not include dashes.

Note: Check payer requirements and format for reporting NDC.

Form Locator 44 HCPCS: Enter the appropriate CPT/HCPCS codes and modifiers, eg,

- Drug: **J2840** for KANUMA® (sebelipase alfa) per 1 mg
- Administration: **96xxx** for drug administration

Note: Some payers may provide specific guidance.

Form Locator 67 Principal Diagnosis Code and 67A–67Q Other Diagnosis Codes: Enter the appropriate diagnosis code, eg,

- ICD-10-CM **E75.5** Other lipid storage disorder

Please see Important Safety Information on pages 1, 9, and 10 and full Prescribing Information for KANUMA® (sebelipase alfa), including Boxed WARNING.

Sample CMS-1450 (UB-04) (or the electronic equivalent 837I): Hospital Outpatient Clinic or Facility[9]

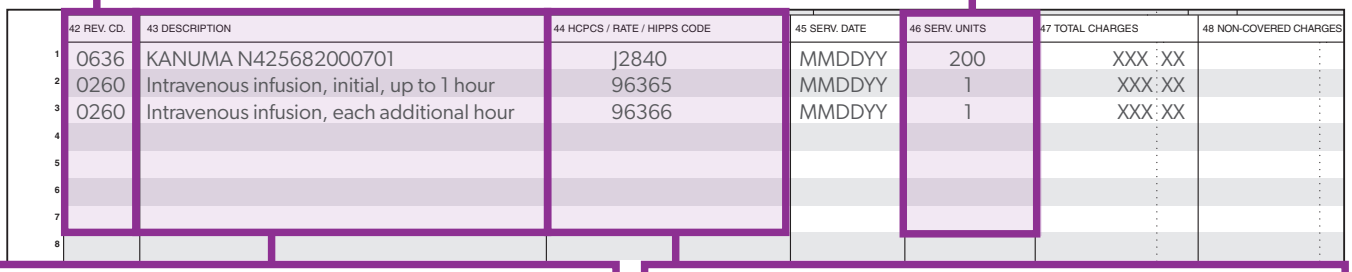
Example claim form for a 200 mg KANUMA® (sebelipase alfa) IV infusion:

Form Locator 42 Revenue Code: Enter the appropriate revenue code,[10] eg,

- **0636** for KANUMA
- **0260** for IV infusion

Note: Other revenue codes may apply.

Form Locator 46 Units of Service: Enter the appropriate number of units of service, eg, KANUMA 200 mg is reported with "200" billing units.



	42 REV. CD.	43 DESCRIPTION	44 HCPCS / RATE / HIPPS CODE	45 SERV. DATE	46 SERV. UNITS	47 TOTAL CHARGES	48 NON-COVERED CHARGES
1	0636	KANUMA N425682000701	J2840	MMDDYY	200	XXX XX	
2	0260	Intravenous infusion, initial, up to 1 hour	96365	MMDDYY	1	XXX XX	
3	0260	Intravenous infusion, each additional hour	96366	MMDDYY	1	XXX XX	
4							
5							
6							
7							
8							

Form Locator 43 Revenue Description: FL 43 Revenue Description: Enter the corresponding drug description for the HCPCS listed in FL 44.

Enter the NDC number followed by the drug name in FL 43.

- The "N4" qualifier is required before the NDC; do not include dashes.

Note: Check payer requirements and format for reporting NDC.

Form Locator 44 HCPCS: Enter the appropriate CPT/HCPCS codes and modifiers, eg,

- Drug: **J2840** for KANUMA® (sebelipase alfa) per 1 mg
- Administration: **96xxx** for drug administration

Note: Some payers may provide specific guidance.

CREATION DATE | TOTALS

52 REL. INFO | 53 ASG. BEN. | 54 PRIOR PAYMENTS | 55 EST. AMOUNT DUE | 56 NPI | 57 OTHER PRV ID

58 INSURED'S NAME | 59 P.REL | 60 INSURED'S UNIQUE ID | 61 GROUP NAME | 62 INSURANCE GROUP NO.

63 TREATMENT AU

Form Locator 67 Principal Diagnosis Code and 67A–67Q Other Diagnosis Codes: Enter the appropriate diagnosis code, eg,

- ICD-10-CM **E75.5** Other lipid storage disorder

66 DX | E75.5 | A I | B J | C K | D L | E M | F N | G O | H P | Q | 68

69 ADMIT DX | 70 PATIENT REASON DX | a | b | c | 71 PPS CODE | 72 ECI | a | b | c | 73

74 PRINCIPAL PROCEDURE CODE DATE | a. OTHER PROCEDURE CODE DATE | b. OTHER PROCEDURE CODE DATE | 75 | 76 ATTENDING | NPI | QUAL

Please see Important Safety Information on pages 1, 9, and 10 and full Prescribing Information for KANUMA® (sebelipase alfa), including Boxed WARNING.

Explore Alexion Access Navigator

ALEXION ACCESS NAVIGATOR

Alexion Access Navigator is a dedicated resource website for US Healthcare Professionals and their offices that contains downloadable access and reimbursement materials for KANUMA® (sebelipase alfa).

Online: https://alexionaccessnavigator.com

OneSource™ OFFERS PATIENT SUPPORT

Contact OneSource:

Phone:
1-888-765-4747
Monday to Friday, 8:30 AM to 8:00 PM ET

Online:
https://alexiononesource.com

IMPORTANT SAFETY INFORMATION INCLUDING BOXED WARNING (cont.)

WARNINGS AND PRECAUTIONS

Hypersensitivity Reactions Including Anaphylaxis

Life-threatening hypersensitivity reactions, including anaphylaxis, have been reported in patients treated with enzyme replacement therapies, including KANUMA. These reactions in KANUMA-treated patients were based on application of Sampson criteria to identify signs/symptoms consistent with anaphylaxis. In clinical trials, 3 (infants) of 106 (3%) patients treated with KANUMA experienced signs and symptoms consistent with anaphylaxis. These patients experienced reactions during infusion with signs and symptoms including chest discomfort, conjunctival injection, dyspnea, generalized and itchy rash, hyperemia, swelling of eyelids, rhinorrhea, severe respiratory distress, tachycardia, tachypnea, and urticaria.

In clinical trials, 21 of 106 (20%) KANUMA-treated patients, including 9 of 14 (64%) infants and 12 of 92 (13%) pediatric patients who were 4 years and older and adults, experienced signs and symptoms either consistent with or that may be related to a hypersensitivity reaction. Signs and symptoms of hypersensitivity reactions, occurring in two or more patients, included abdominal pain, agitation, fever, chills, diarrhea, eczema, edema, hypertension, irritability, laryngeal edema, nausea, pallor, pruritus, rash, and vomiting. Most reactions occurred during or within 4 hours of the completion of the infusion. Patients were not routinely pre-medicated prior to infusion of KANUMA in these clinical trials.

Administration of KANUMA should be supervised by a healthcare provider knowledgeable in the management of severe hypersensitivity reactions including anaphylaxis. Observe patients closely during and after the infusion.

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IMPORTANT SAFETY INFORMATION INCLUDING BOXED WARNING (cont.)

WARNINGS AND PRECAUTIONS (cont.)

Hypersensitivity Reactions Including Anaphylaxis (cont.)

The management of hypersensitivity reactions should be based on the severity of the reaction and may include temporarily interrupting the infusion, lowering the infusion rate, and/or treatment with antihistamines, antipyretics, and/or corticosteroids. If interrupted, the infusion may be resumed at a slower rate with increases as tolerated. Pre-treatment with antipyretics and/or antihistamines may prevent subsequent reactions in those cases where symptomatic treatment was required.

Consider the risks and benefits of re-administering KANUMA following a severe reaction. Monitor patients, with appropriate resuscitation measures available, if the decision is made to re-administer the product.

Hypersensitivity to Eggs or Egg Products

Patients with a known history of egg allergies were excluded from the clinical trials. Consider the risks and benefits of treatment with KANUMA in patients with known systemic hypersensitivity reactions to eggs or egg products.

ADVERSE REACTIONS

In clinical trials, the most common adverse reactions were:

- Infants with Rapidly Progressive LAL Deficiency Presenting within the First 6 Months of Life (≥30%): diarrhea, vomiting, fever, rhinitis, anemia, cough, nasopharyngitis, and urticaria.
- Pediatric and Adult Patients with LAL Deficiency (≥8%): headache, fever, oropharyngeal pain, nasopharyngitis, asthenia, constipation, and nausea.

To report SUSPECTED ADVERSE REACTIONS, contact Alexion Pharmaceuticals, Inc. at 1-844-259-6783 or FDA at 1-800-FDA-1088 or www.fda.gov/medwatch

REFERENCES

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ALEXION®
AstraZeneca Rare Disease