Hizentra®
Immune Globulin Subcutaneous (Human) 20% Liquid

Coding Information for Primary Immune Deficiency (PI)

Diagnosis and ICD-10-CM billing codes for 20% subcutaneous Ig (SCIg) therapy

Biotherapies for Life®

CSL Behring

Please see full Important Safety Information on back and enclosed full prescribing information for Hizentra, including boxed warning.

Hizentra Coding Information

Diagnosis and billing codes for 20% SCIg therapy

This resource provides information 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[1] that describe the patient's condition and are supported by the medical record. All codes listed in this guide are for informational purposes and are not an exhaustive list. The CPT®,[2] HCPCS,[3] and ICD-10-CM 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.

Medicare, Medicaid, and most insurers cover Hizentra for the treatment of patients with primary immunodeficiency (PI) disease. Hizentra is covered as a Medicare Part B benefit with claims considered for payment by regional Durable Medical Equipment Medicare Administrative Contractors (DME MACs).

Medicaid coverage policy varies by state. Coverage by other payers varies by payer and provider contract.

ICD-10-CM Diagnosis Codes[1]

The following ICD-10-CM codes[1] may be used to identify patient immunologic medical conditions typically associated with Hizentra use.

D80 Immunodeficiency with predominantly antibody defects

- D80.0* Hereditary hypogammaglobulinemia
  Autosomal recessive agammaglobulinemia (Swiss type)
  X-linked agammaglobulinemia [Bruton]
  (with growth hormone deficiency)
- D80.1 Nonfamilial hypogammaglobulinemia
  Agammaglobulinemia with immunoglobulin-bearing B-lymphocytes
  Common variable agammaglobulinemia [CVAgamma]
  Hypogammaglobulinemia NOS
- D80.2* Selective deficiency of immunoglobulin A [IgA]
- D80.3* Selective deficiency of immunoglobulin G [IgG] subclasses
- D80.4* Selective deficiency of immunoglobulin M [IgM]
- D80.5* Immunodeficiency with increased immunoglobulin M [IgM]
- D80.6* Antibody deficiency with near-normal immunoglobulins or with hyperimmunoglobulinemia
- D80.7* Transient hypogammaglobulinemia of infancy
- D80.8 Other immunodeficiencies with predominantly antibody defects
  Kappa light chain deficiency
- D80.9 Immunodeficiency with predominantly antibody defects, unspecified

D81 Combined immunodeficiencies

- D81.0* Severe combined immunodeficiency [SCID] with reticular dysgenesis
- D81.1* Severe combined immunodeficiency [SCID] with low T- and B-cell numbers
- D81.2* Severe combined immunodeficiency [SCID] with low or normal B-cell numbers
- D81.4 Nezelof's syndrome
- D81.5* Purine nucleoside phosphorylase [PNP] deficiency
- D81.6* Major histocompatibility complex class I deficiency
  Bare lymphocyte syndrome
- D81.7* Major histocompatibility complex class II deficiency
- D81.89* Other combined immunodeficiencies
- D81.9* Combined immunodeficiency, unspecified
  Severe combined immunodeficiency disorder [SCID] NOS

D82 Immunodeficiency associated with other major defects

Excludes: ataxia telangiectasia [Louis-Bar] (G11.3)

- D82.0* Wiskott-Aldrich syndrome
  Immunodeficiency with thrombocytopenia and eczema
- D82.1* Di George's syndrome
  Pharyngeal pouch syndrome
  Thymic alymphoplasia
  Thymic aplasia or hypoplasia with immunodeficiency
- D82.2 Immunodeficiency with short-limbed stature
- D82.3 Immunodeficiency following hereditary defective response to Epstein-Barr virus
  X-linked lymphoproliferative disease
- D82.4* Hyperimmunoglobulin E [IgE] syndrome
- D82.8 Immunodeficiency associated with other specified major defects
- D82.9 Immunodeficiency associated with major defect, unspecified

D83 Common variable immunodeficiency

- D83.0* Common variable immunodeficiency with predominant abnormalities of B-cell numbers and function
- D83.1* Common variable immunodeficiency with predominant immunoregulatory T-cell disorders
- D83.2* Common variable immunodeficiency with autoantibodies to B- or T-cells
- D83.8* Other common variable immunodeficiencies
- D83.9* Common variable immunodeficiency, unspecified

D84 Immunodeficiency, unspecified

- D84.9 Immunodeficiency, unspecified

G11 Hereditary Ataxia

- G11.3* Cerebellar ataxia with defective DNA repair
  Ataxia telangiectasia

*Medicare Part B–approved diagnosis codes for treatment with Hizentra in the home. All other diagnoses may qualify for coverage under Medicare Part D plans.

ICD=International Classification of Diseases
HCPCS=Healthcare Common Procedure Coding System
CPT=Current Procedural Terminology

Prior Authorization

When a payer requires prior authorization:

- Request the approval and authorization reference code in writing
- Insert the authorization reference code into Field 23 of the CMS-1500 claim form when billing for services. Enter the authorization reference code in the equivalent field on the electronic claim form (ASC 837P, Loop 2300, Segment REF02, with the Qualifier G1 entered to Segment REF01), unless otherwise directed by the payer
- If required by the payer, attach a copy of the authorization approval letter to the paper claim form; use the payer's designated procedure for submitting documentation with electronic claim forms

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Billing Codes

The following HCPCS codes describe supplies (including drugs) rendered by the billing provider. CMS publishes and maintains the HCPCS code set.

These codes are entered to paper claim form CMS-1500 in Field 24; or, to electronic claim form ASC 837P in Loop 2400, Segment SV101-2, with the Qualifier "HC" entered to Segment SV101-1, unless otherwise directed by the payer.

Hizentra, Immune Globulin Subcutaneous (Human), 20% Liquid[3]

HCPCS Code	Description
J1559	Injection, immune globulin (Hizentra), 100 mg

Certain payers may require the entry of NDC information to the claim form, as directed by the payer.

Ready-to-Use Hizentra Vial Sizes

Grams Protein	Fill Size	NDC Number to Use on All Claim Forms
1 g	5 mL	44206-0451-01
2 g	10 mL	44206-0452-02
4 g	20 mL	44206-0454-04
10 g	50 mL	44206-0455-10

Ready-to-Use Hizentra Prefilled Syringe Sizes

Grams Protein	Fill Size	NDC Number to Use on All Claim Forms
1 g	5 mL	44206-456-21
2 g	10 mL	44206-457-22
4 g	20 mL	44206-458-24

If the billing provider performs administration services in conjunction with a patient's infusion, the following administration codes may be used to bill for this service if the service meets the requirements of the code description.

CPT Code[2]	Description
96369	Subcutaneous injection for therapy or prophylaxis; initial, up to one hour, including pump setup and establishment of subcutaneous infusion site(s)
96370	Each additional hour
96371	Additional pump setup with establishment of new subcutaneous infusion site(s)

Billing for Medicare DME MACs[3]

The Medicare benefit for subcutaneous immune globulin administered by Durable Medical Equipment Medicare Administrative Contractors (DME MACs) applies only to those products that are specifically labeled as subcutaneous administration products. Intravenous immune globulin products administered via the subcutaneous route are not covered by Medicare DME MACs (refer to Local Coverage Decision for External Infusion Pumps, effective 2/4/2011).

Billing for External Pumps and Supplies

When the billing provider furnishes an external infusion pump for patient use, the following code may be used to bill for the pump if it meets the requirements of the code description.

External Infusion Pump

HCPCS Code	Description
E0779	Ambulatory infusion pump, mechanical, reusable, for infusion 8 hours or greater

E0779 is the **only** reimbursable pump code for infusion of Hizentra under Medicare Part B.

Other pump codes may be used to bill other payers, depending on the pump that is used and the payer's pump coverage policy. The codes below are not covered by Medicare. Other pump codes may include:

E0781	Ambulatory infusion pump, single or multiple channels, electric or battery operated, with administrative equipment, worn by patient
E0791	Parenteral infusion pump, stationary, single or multichannel

When the billing provider furnishes supplies for the patient's use with the external infusion pump, the following codes may be appropriate for the supplies if they meet the requirements of the code description and payer coverage policies.

Professional Services for Drug Infusion

G0069	Professional services for the administration of subcutaneous immunotherapy for each infusion drug administration calendar day in the individual's home, each 15 minutes

External Infusion Pump Supplies*

K0552	Supplies for external drug infusion pump, syringe-type cartridge, sterile, each
A4221	Supplies for maintenance of drug infusion catheter, per week
A4222	Infusion supplies for external drug infusion pump, per cassette or bag

*All 3 codes are needed to bill for infusion pump supplies.

Modifier -JB for Medicare Billing

Medicare requires the addition of Modifier -JB (administered subcutaneously) to the HCPCS code for Hizentra (J1559) to confirm subcutaneous administration. Example: J1559-JB.
JB should also be added to the HCPCS code for the covered infusion pump (E0779) following the modifier that indicates whether the pump is being rented (RR) or purchased (NU). Example: E0779-RR-JB or E0779-NU-JB.

A Field 21

Enter all appropriate ICD-10-CM diagnosis codes, starting on Field 21, Line A. This field allows the entry of 1 character indicator and 12 diagnosis codes at a maximum of 7 characters in length.

B Field 24D (CPT/HCPCS)

- Enter HCPCS code J1559 for Hizentra if appropriate
- Enter HCPCS codes for pump and related supplies
  - Pump: Enter appropriate code (Medicare will cover only E0779)
  - Supplies: Enter code that best describes supplies provided (eg, K0552 or A4221)
- Enter G0069 for Medicare Claims each day when services were provided in the home
- If the billing provider is eligible for coverage of subcutaneous infusion, enter CPT code 96369 for the establishment of infusion sites, pump setup, and the first hour of infusion. If the patient is monitored beyond the first hour, CPT code 96370 may be appropriate. Additional pump setup with establishment of new subcutaneous infusion sites is reported with CPT code 96371

C Field 24D (Shaded Area)

For Medicaid claims, and for Medicare claims that will cross over to Medicaid as the secondary payer, NDC information in a specific format is required in the shaded area above the line on which Hizentra is reported in 24D. **The various Medicaid plans and Medicare have different reporting formats for this information.** In general, the billing entity will need to supply the NDC (in HIPAA-compliant 11-digit format) preceded by the Modifier N4 (eg, N499999999999). This is typically followed by the NDC unit of measure (F2 [international unit], GR [gram], ML [milliliter], or UN [unit]) and the numeric quantity of the NDC that was dispensed.

Other payers may require similar information. Check with your payer for specific requirements related to reporting the information required in the shaded areas of Field 24.

D Field 24D (Modifier)

For Medicare claims, enter Modifier -JB (administered subcutaneously) on the same line as Hizentra (ie, J1559-JB). Modifier -JB must also be added to the line for the pump following the modifier that indicates whether the pump is a rental (RR) or a purchase (NU) (eg, E0779-RR-JB or E0779-NU-JB).

E Field 24E (Diagnosis Pointer)

Enter the line letter(s) from Field 21 that best describes the medical necessity for the service listed in Field 24D. For Medicare claims, only one line number from Field 21 should be entered in Field 24E for each HCPCS code reported in Field 24D.

F Field 24G (Days or Units)

- Hizentra: Enter the number of 100-mg billing units used to treat the patient. For example, if 40,000 mg are dispensed (eg, four 50-mL vials), 400 would be reported in Field 24G (40,000/100 = 400)
- Pump: Enter one billing unit
- Supplies: Enter one per unit described by the corresponding HCPCS code

CMS-1500 Claim Example

HEALTH INSURANCE CLAIM FORM

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

PICA | PICA

1. MEDICARE (Medicare#) | MEDICAID (Medicaid#) | TRICARE (ID#/DoD#) | CHAMPVA (Member ID#) | GROUP HEALTH PLAN (ID#) | FECA BLK LUNG (ID#) | OTHER (ID#)

1a. INSURED'S I.D. 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 F

4. INSURED'S NAME (Last Name, First Name, Middle Initial)

5. PATIENT'S ADDRESS (No., Street)

6. PATIENT RELATIONSHIP TO INSURED Self | Spouse | Child | Other

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) YES NO

a. INSURED'S DATE OF BIRTH MM DD YY SEX M F

b. RESERVED FOR NUCC USE

b. AUTO ACCIDENT? YES NO PLACE (State)

b. OTHER CLAIM ID (Designated by NUCC)

c. RESERVED FOR NUCC USE

c. OTHER ACCIDENT? YES 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? YES NO *If yes*, complete items 9, 9a, and 9d.

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.

SIGNED _____ DATE _____

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 _____

14. DATE OF CURRENT ILLNESS, INJURY, or PREGNANCY (LMP) MM DD YY QUAL.

15. OTHER DATE QUAL. MM DD YY

16. DATES PATIENT UNABLE TO WORK IN CURRENT OCCUPATION MM DD YY FROM MM DD YY TO

17. NAME OF REFERRING PROVIDER OR OTHER SOURCE

17a.

17b. NPI

18. HOSPITALIZATION DATES RELATED TO CURRENT SERVICES MM DD YY FROM MM DD YY TO

19. ADDITIONAL CLAIM INFORMATION (Designated by NUCC)

20. OUTSIDE LAB? YES NO $ CHARGES

21. DIAGNOSIS OR NATURE OF ILLNESS OR INJURY Relate A-L to service line below (24E) ICD Ind.

A. B. C. D.

E. F. G. H.

I. J. K. L.

22. RESUBMISSION CODE ORIGINAL REF. NO.

23. PRIOR AUTHORIZATION NUMBER

24. A. DATE(S) OF SERVICE From MM DD YY	To MM DD YY	B. PLACE OF SERVICE	C. EMG	D. PROCEDURES, SERVICES, OR SUPPLIES (Explain Unusual Circumstances) CPT/HCPCS	MODIFIER	E. DIAGNOSIS POINTER	F. $ CHARGES	G. DAYS OR UNITS	H. EPSDT Family Plan	I. ID. QUAL.	J. RENDERING PROVIDER ID. #
1										NPI	
2										NPI	
3										NPI	
4										NPI	
5										NPI	
6										NPI	

25. FEDERAL TAX I.D. NUMBER SSN EIN

26. PATIENT'S ACCOUNT NO.

27. ACCEPT ASSIGNMENT? (For govt. claims, see back) YES NO

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.)

SIGNED DATE

32. SERVICE FACILITY LOCATION INFORMATION

a. NPI b.

33. BILLING PROVIDER INFO & PH # ( )

a. NPI b.

CARRIER

PATIENT AND INSURED INFORMATION

PHYSICIAN OR SUPPLIER INFORMATION

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

Call Us Now for Reimbursement Support

Hizentra reimbursement support is available free of charge through **IgIQ®**, CSL Behring's single source for Ig solutions.

IgIQ® Your single source for Ig solutions

1-877-355-IGIQ (4447)

Monday—Friday 8 AM to 8 PM ET

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). 2020 Codes Tables and Index. https://www.cdc.gov/nchs/icd/icd10cm.htm. Accessed January 30, 2020. **2.** American Medical Association. Medicine/hydration, therapeutic, prophylactic, diagnostic injections and infusions, and chemotherapy. In: American Medical Association, ed. *CPT 2016*. Chicago, IL: American Medical Association; 2016:650-651. **3.** Centers for Medicare and Medicaid Services (US Department of Health and Human Services). HCPCS Release & Code Sets. https://www.cms.gov/Medicare/Coding/HCPCSReleaseCodeSets/Alpha-Numeric-HCPCS. Accessed January 30, 2020.

Important Safety Information

Hizentra is indicated for:

- Treatment of primary immunodeficiency (PI) in adults and pediatric patients 2 years and older.
- Maintenance therapy in adults with chronic inflammatory demyelinating polyneuropathy (CIDP) to prevent relapse of neuromuscular disability and impairment.
  - Limitation of Use: Maintenance therapy in CIDP has been systematically studied for 6 months and for a further 12 months in a follow-up study. Continued maintenance beyond these periods should be individualized based on patient response and need for continued therapy.

For subcutaneous infusion only.

- **WARNING: Thrombosis may occur with immune globulin products, including Hizentra. Risk factors may include: advanced age, prolonged immobilization, hypercoagulable conditions, history of venous or arterial thrombosis, use of estrogens, indwelling vascular catheters, hyperviscosity, and cardiovascular risk factors.**
- **For patients at risk of thrombosis, administer Hizentra at the minimum dose and infusion rate practicable. Ensure adequate hydration in patients before administration. Monitor for signs and symptoms of thrombosis and assess blood viscosity in patients at risk for hyperviscosity.**

Hizentra is contraindicated in patients with a history of anaphylactic or severe systemic reaction to human immune globulin (Ig) or components of Hizentra (eg, polysorbate 80), as well as in patients with immunoglobulin A deficiency with antibodies against IgA and a history of hypersensitivity. Because Hizentra contains L-proline as stabilizer, use in patients with hyperprolinemia is contraindicated.

IgA-deficient patients with anti-IgA antibodies are at greater risk of severe hypersensitivity and anaphylactic reactions. Thrombosis may occur following treatment with Ig products, including Hizentra.

Monitor patients for aseptic meningitis syndrome (AMS), which may occur following treatment with Ig products, including Hizentra. In patients at risk of acute renal failure, monitor renal function, including blood urea nitrogen, serum creatinine and urine output. In addition, monitor patients for clinical signs of hemolysis or pulmonary adverse reactions (eg, transfusion-related acute lung injury [TRALI]).

Hizentra is derived from human blood. 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.

The most common adverse reactions (observed in ≥5% of study subjects) were local infusion-site reactions, as well as headache, diarrhea, fatigue, back pain, nausea, extremity pain, cough, upper respiratory tract infection, rash, pruritus, vomiting, upper abdominal pain, migraine, arthralgia, pain, fall, and nasopharyngitis.

The passive transfer of antibodies can interfere with response to live virus vaccines and lead to misinterpretation of serologic test results.

Please see enclosed full prescribing information for Hizentra.

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.

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