



## Drug Coverage Policy

Effective Date ..... 12/15/2025

Coverage Policy Number.....IP0555

Policy Title.....Vonvendi

# Hematology – Vonvendi

- Vonvendi® (von Willebrand factor [recombinant] intravenous infusion – Baxalta)

---

### 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. Each coverage request should be reviewed on its own merits. Medical directors are expected to exercise clinical judgment where appropriate and have discretion in making individual coverage determinations. Where coverage for care or services does not depend on specific circumstances, reimbursement will only be provided if a requested service(s) is submitted in accordance with the relevant criteria outlined in the applicable Coverage Policy, including covered diagnosis and/or procedure code(s). Reimbursement is not allowed for services when billed for conditions or diagnoses that are not covered under this Coverage Policy (see "Coding Information" below). When billing, providers must use the most appropriate codes as of the effective date of the submission. Claims submitted for services that are not accompanied by covered code(s) under the applicable Coverage Policy will be denied as not covered. 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

Vonvendi, a recombinant von Willebrand factor (VWF), is indicated for **routine prophylaxis in von Willebrand disease (VWD)** to reduce the frequency of bleeding episodes in adults.

Vonvendi is also indicated for use in **VWD in adult and pediatric patients** for:<sup>1</sup>

- **On-demand treatment and control** of bleeding episodes.
- **Perioperative management** of bleeding.

## Disease Overview

VWD is an inherited bleeding disorder caused by a deficiency or impairment of a protein found in blood called VWF.<sup>2-4</sup> VWF is a plasma protein with a dual role in hemostasis by mediating platelet adhesion at sites of vascular injury and by binding and stabilizing Factor VIII. The disease is rather common as it affects 1 in 100 people; both genders are impacted equally. Symptoms of VWD include mucocutaneous bleeding and excessive hemorrhage following invasive procedures; occasionally, soft tissue hematomas and joint bleeding may also occur. Women who have VWD may experience heavy menorrhagia or experience excessive bleeding at childbirth. Bleeding episodes may be life-threatening in patients with severe forms of VWD. VWD is classified into six types (1, 2A, 2B, 2M, 2N, and 3) according to distinct genotypic, clinical, and laboratory phenotypic characteristics. Type 1 VWD is the most common type (60% to 80% of patients) and represents a partial quantitative deficiency of VWF. Bleeding symptoms are generally mild to moderate. Type 2 VWD affects 15% to 30% of patients and consists of four disease subtypes (2A, 2B, 2M, and 2N) dependent on the specific gene mutation (e.g., decreased VWF-dependent platelet adhesion, decreased binding affinity for Factor VIII). This type is due to a qualitative VWF defect, and the bleeding is generally moderate, but can vary among patients. Type 3 VWD is uncommon (5% to 10% of patients) but is usually severe because it is due to a virtually complete deficiency of VWF. Many patients with VWD also have reduced Factor VIII levels. Treatment options for VWD include desmopressin either parenterally or by a highly concentrated nasal spray (Stimate), Vonvendi, or plasma-derived Factor VIII product that contain VWF.

## Guidelines

The National Bleeding Disorders Foundation Medical and Scientific Advisory Council has guidelines for the treatment of hemophilia and other bleeding disorders (revised October 2024).<sup>4</sup> Most patients with Type 1 VWD may be treated with a desmopressin product (DDAVP injection or Stimate nasal spray). Some patients with type 2A VWD may respond to DDAVP; a clinical trial with DDAVP should be performed to determine if DDAVP can be used for these particular patients. The guidelines recommend that both DDAVP injection and Stimate not be used in children aged < 2 years and in patients with VWD in whom desmopressin does not provide adequate VWF levels. Also, they should be used cautiously in pregnant women during labor and delivery. Use of plasma-derived VWF-containing Factor VIII concentrates that have VWF is recommended in certain types of VWD that do not respond to therapy with desmopressin (i.e., Type 2B VWD and Type 3 VWD). Also, plasma-derived Factor VIII concentrates that contain VWF (e.g., Alphanate, Humate-P, and Wilate) are recommended in Types 1, 2A, 2M, and 2N VWD who have become transiently unresponsive to DDAVP, as well as in surgical situations, especially in young children < 2 years of age. Wilate is FDA-approved for routine prophylaxis in children ≥ 6 years of age with VWD. Cryoprecipitate should not be utilized to treat patients with VWD except in life- and limb-threatening emergencies when VWD-containing factor VIII concentrate is not immediately available. Vonvendi is available to treat patients with Type 2B and Type 3 VWD; it can also be used in patients with Types 1, 2A, 2M, and 2N VWD who are not responsive to DDAVP and in children < 2 years of age, regardless of VWD type. Vonvendi is approved for use as routine prophylaxis only in patients with severe Type 3 VWD who were previously treated with VWF (recombinant or plasma-derived) on demand. It is produced in Chinese hamster ovary cells and it does not contain human or animal-derived proteins in its cell culture or in its final formulation (a third generation product). Vonvendi contains ultra-large VWF multimers, in addition to the high, medium, and low molecular weight VWF multimers normally found in plasma. Trace amounts of recombinant Factor VIII is in the product as well.

## Dosing Considerations

Dosing of clotting factor concentrates is highly individualized. MASAC provides recommendations regarding doses of clotting factor concentrate in the home (2016).<sup>5</sup> The number of required doses varies greatly and is dependent on the severity of the disorder and the prescribed regimen. Per MASAC guidance, patients on prophylaxis should also have a minimum of one major dose and two

minor doses on hand for breakthrough bleeding in addition to the prophylactic doses used monthly. The guidance also notes that an adequate supply of clotting factor concentrate is needed to accommodate weekends and holidays. Therefore, maximum doses in this policy allow for prophylactic dosing plus three days of acute bleeding or perioperative management per 28 days. Doses exceeding this quantity will be reviewed on a case-by-case basis by a clinician.

## Coverage Policy

### POLICY STATEMENT

Prior Authorization is required for benefit coverage Vonvendi. Approval is recommended for those who meet the **Criteria** and **Dosing** for the listed indication. Extended approvals are allowed if the patient continues to meet the Criteria and Dosing. Requests for doses outside of the established dosing documented in this policy will be considered on a case-by-case basis by a clinician (i.e., Medical Director or Pharmacist). Because of the specialized skills required for evaluation and diagnosis of patients treated with Vonvendi, as well as the monitoring required for adverse events and long-term efficacy, the agent is required to be prescribed by or in consultation with a physician who specializes in the condition being treated.

**Vonvendi is considered medically necessary when the following criteria are met:**

### FDA-Approved Indication

1. **Von Willebrand Disease.** Approve for 1 year if the agent is prescribed by or in consultation with a hematologist.

**Dosing.** Approve the following dosing regimens (A, B, and/or C):

- A)** On demand treatment and control of bleeding episodes: approve up to 900 IU/kg intravenously no more frequently than once every 28 days; AND/OR
- B)** Perioperative management: approve up to 900 IU/kg intravenously no more frequently than once every 28 days; AND/OR
- C)** Routine prophylaxis: approve up to 60 IU/kg intravenously no more frequently than twice weekly.

### Conditions Not Covered

**Vonvendi for any other use is considered not medically necessary. Criteria will be updated as new published data are available.**

## Coding 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
J7179	Injection, von Willebrand factor (recombinant), (Vonvendi), 1 IU VWF:RCo

## References

1. Vonvendi® intravenous infusion [prescribing information]. Cambridge, MA; Takeda; September 2025.
2. National Bleeding Disorders Foundation. MASAC (Medical and Scientific Advisory Council) recommendations concerning products licensed for the treatment of hemophilia and selected disorders of the coagulation system (October 2024). MASAC Document #290. Available at: <https://www.hemophilia.org/sites/default/files/document/files/MASAC-Products-Licensed.pdf>. Accessed on September 12, 2025.
3. Srivastava A, Santagostino E, Dougall A, et al, on behalf of the WFH guidelines for the management of hemophilia panelists and coauthors. WFH guidelines for the management of hemophilia, 3<sup>rd</sup> edition. *Haemophilia*. 2020;26 Suppl 6:1-158.
4. Connell NT, Flood VH, Brignardello-Petersen R, et al. ASH ISTH NHF WFH 2021 guidelines on the management of von Willebrand Disease. *Blood Adv*. 2021;5(1):301-325.
5. National Hemophilia Foundation. MASAC (Medical and Scientific Advisory Council) recommendations regarding doses of clotting factor concentrate in the home (Revised June 7, 2016). MASAC Document #242. Adopted on September 3, 2020. Available at: <https://www.hemophilia.org/sites/default/files/document/files/242.pdf>. Accessed on September 12, 2025.

## Revision Details

Type of Revision	Summary of Changes	Date
Annual Revision	No criteria changes	5/1/2024
Annual Revision	No criteria changes	3/15/2025
Selected Revision	Von Willebrand Disease: Removed age restriction.	12/15/2025

The policy effective date is in force until updated or retired.

---

"Cigna Companies" refers to operating subsidiaries of The Cigna Group. All products and services are provided exclusively by or through such operating subsidiaries, including Cigna Health and Life Insurance Company, Connecticut General Life Insurance Company, Evernorth Behavioral Health, Inc., Cigna Health Management, Inc., and HMO or service company subsidiaries of The Cigna Group. © 2025 The Cigna Group.