

<b>Reference number(s)</b>
1951-A

## SPECIALTY GUIDELINE MANAGEMENT

### VONVENDI (von Willebrand factor [recombinant])

#### POLICY

##### I. INDICATIONS

The indications below including FDA-approved indications and compendial uses are considered a covered benefit provided that all the approval criteria are met and the member has no exclusions to the prescribed therapy.

##### FDA-Approved Indication

Vonvendi is indicated for use in adults (age 18 and older) diagnosed with von Willebrand disease (VWD) for:

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

All other indications are considered experimental/investigational and not medically necessary.

##### II. CRITERIA FOR INITIAL APPROVAL

##### **Von Willebrand Disease**

Authorization of 12 months may be granted for treatment of VWD when any of the following criteria is met:

- A. Member has type 1, 2A, 2M, or 2N VWD and has had an insufficient response to desmopressin or a documented clinical reason for not using desmopressin (see Appendix).
- B. Member has type 2B or type 3 VWD.

##### III. CONTINUATION OF THERAPY

Authorization of 12 months may be granted for continued treatment in members requesting reauthorization for an indication listed in Section II when the member is experiencing benefit from therapy (e.g., reduced frequency or severity of bleeds).

##### IV. APPENDIX

##### **Clinical Reasons for Not Utilizing Desmopressin in Patients with Type 1, 2A, 2N and 2M VWD**

- A. Age < 2 years
- B. Pregnancy
- C. Fluid/electrolyte imbalance
- D. High risk for cardiovascular or cerebrovascular disease (especially the elderly)
- E. Predisposition to thrombus formation
- F. Trauma requiring surgery
- G. Life-threatening bleed
- H. Contraindication or intolerance to desmopressin
- I. Severe type 1 von Willebrand disease

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## V. REFERENCES

1. Vonvendi [package insert]. Lexington, MA: Baxalta US Inc.; February 2019.
2. National Hemophilia Foundation. MASAC recommendations concerning products licensed for the treatment of hemophilia and other bleeding disorders. Revised August 2020. MASAC Document #263. [https://www.hemophilia.org/sites/default/files/document/files/263\\_treatment.pdf](https://www.hemophilia.org/sites/default/files/document/files/263_treatment.pdf). Accessed October 14, 2020.
3. National Hemophilia Foundation. MASAC recommendations regarding the treatment of von Willebrand disease. Revised November 2016. MASAC Document #244. <https://www.hemophilia.org/sites/default/files/document/files/244.pdf>. Accessed October 14, 2020.
4. National Institutes of Health. The diagnosis, evaluation, and management of von Willebrand disease. Bethesda, MD: US Dept of Health and Human Services, National Institutes of Health; 2007. NIH publication No. 08-5832.
5. Stimate [package insert]. King of Prussia, PA: CSL Behring LLC; June 2013.
6. Leissinger C, Carcao M, Gill JC, et al. Desmopressin (DDAVP) in the management of patients with congenital bleeding disorders. *Haemophilia*. 2014;20:158-167.