

<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

On-demand treatment and control of bleeding episodes in adults with von Willebrand disease (vWD)

All other indications are considered experimental/investigational and are not a covered benefit.

##### II. CRITERIA FOR INITIAL APPROVAL

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

12 months of authorization 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

All members (including new members) requesting authorization for continuation of therapy must meet ALL initial authorization criteria.

##### 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

##### V. REFERENCES

1. Vonvendi [package insert]. Westlake Village, CA: Baxalta US Inc.; December 2015.

Reference number(s)
1951-A

2. National Hemophilia Foundation. MASAC recommendations concerning products licensed for the treatment of hemophilia and other bleeding disorders. Revised August 2017. MASAC Document # 250. Accessed December 8, 2017.
3. National Hemophilia Foundation. MASAC recommendations regarding the treatment of von Willebrand disease. Revised November 2016. MASAC Document #244. Accessed December 8, 2017.
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. Leissing C, Carcao M, Gill JC, et al. Desmopressin (DDAVP) in the management of patients with congenital bleeding disorders. *Haemophilia*. 2014;20:158-167.