Emergency Care for Patients with von Willebrand Disease

Authors and Editors: Susan C. Zappa RN, Lucie Lacasse RN, Rose Jacobson RN, Sherry Purcell RN and Karen Wulff RN
Medical Reviewers: David Lillicrap MD, FRCPC and Marcela Torres MD

von Willebrand Disease (VWD) is classified by ‘type 1, 2, or 3’ If the type is unknown proceed as if type 1, if bleeding continues consult a hematologist.

Mucous membrane bleeds (pg. 6) Administer the recommended treatment and anti-fibrinolytics

Gynecological bleeds (pg. 12)

Immobilizers p.r.n. for joint bleeds

Abdominal bleeds (pg. 10)

Trauma (pg. 23) Administer the recommended treatment

For VWD type 3: Avoid intra-muscular injections due to the possibility of causing a muscle bleed

Minor cuts / bruises no treatment

Head Injury (pg. 4) Always treat immediately with the recommended treatment

Ice pack for soft tissue, muscle, joint bleeds
# Treatment and Management Guidelines for von Willebrand Disease

<table>
<thead>
<tr>
<th>Type of von Willebrand Disease</th>
<th>Major life-threatening bleeds</th>
<th>Other bleeds</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>(ex. - head injury, GI bleeding, severe menorrhagia, etc.)</td>
<td>(ex. - sutures, nosebleed, mouth bleed, dental extractions etc.)</td>
</tr>
<tr>
<td>Type 1 or Type 2</td>
<td>Factor concentrate containing both FVIII (8) and von Willebrand factor (eg. Humate P®, Alphanate®, wilate®): 60-80 Ristocetin cofactor units/kg IV</td>
<td>Known to respond to desmopressin (DDAVP®): Desmopressin 0.3 mcg/kg IV in 50 ml of Normal Saline over 30 minutes or subcutaneously if volume can be given safely. Recommendation: a maximum dose of 20 mcg. Mucosal bleeding - anti-fibrinolytics (pg. 9) For patients who do not respond to desmopressin: Give a factor concentrate containing both FVIII (8) and von Willebrand factor (eg. Humate P®, Alphanate®, wilate®): 40-60 Ristocetin cofactor units/kg IV Package insert will instruct as to rate per volume. Note: monoclonal or recombinant factor VIII (8) products do NOT have von Willebrand factor in them and will not stop the bleeding.</td>
</tr>
<tr>
<td>The type 2 VWD known as ‘pseudo VWD or VWF platelet type’ will only respond to a platelet transfusion - call a hematologist.</td>
<td>Package insert will instruct as to rate per volume.</td>
<td></td>
</tr>
<tr>
<td>Type 3</td>
<td>Factor concentrate containing both FVIII (8) and von Willebrand factor (eg. Humate P®, Alphanate®, wilate®): 60-80 Ristocetin cofactor units/kg</td>
<td>Factor concentrate containing both FVIII (8) and von Willebrand factor (eg. Humate P®, Alphanate®, wilate®): 40-60 Ristocetin cofactor units/kg</td>
</tr>
<tr>
<td>Most severe form of VWD.</td>
<td>Package insert will instruct as to rate per volume. Note: monoclonal or recombinant factor VIII (8) products do NOT have von Willebrand factor in them and will not stop the bleeding.</td>
<td>Package insert will instruct as to rate per volume. Note: monoclonal or recombinant factor VIII (8) products do NOT have von Willebrand factor in them and will not stop the bleeding.</td>
</tr>
</tbody>
</table>

If your institution does not have Humate-P®, or Alphanate®, wilate®, but does have Koate DVI® available, consult a hematologist for guidelines and instructions.

Per the Medical and Scientific Advisory Council of the National Hemophilia Foundation:
Because of the increased risk of HIV and hepatitis A, B, and C transmission, cryoprecipitate should not be used (for the treatment of von Willebrand Disease) except in an emergency situation where one of the above products is not available and delay of treatment would be life or limb threatening.
<table>
<thead>
<tr>
<th>Province</th>
<th>Region / Program Name</th>
<th>Location</th>
<th>Contact Information</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>BRITISH COLUMBIA</strong></td>
<td>Hemophilia Program of BC (Adult Division)</td>
<td>Vancouver, BC</td>
<td>Tel: (604) 806-8855 / 1-877-806-8855 After hours: (604) 682-2344</td>
</tr>
<tr>
<td></td>
<td>Pediatric Hemophilia / Hematology</td>
<td>Vancouver, BC</td>
<td>Tel: (604) 875-2345 ext. 5335 After hours: (604) 875-2161</td>
</tr>
<tr>
<td></td>
<td>BC Children’s Hospital</td>
<td>Vancouver, BC</td>
<td>Tel: (604) 875-2161 After hours: (604) 875-2161</td>
</tr>
<tr>
<td><strong>ALBERTA</strong></td>
<td>Southern Alberta Hemophilia Program</td>
<td>Calgary, AB</td>
<td>Tel: (403) 955-7311 After hours: (403) 955-7070</td>
</tr>
<tr>
<td></td>
<td>Alberta Children’s Hospital</td>
<td>Calgary, AB</td>
<td>Tel: (403) 944-4057 After hours: (403) 944-1110</td>
</tr>
<tr>
<td></td>
<td>Southern Alberta Rare Blood &amp; Bleeding Disorders (Adults)</td>
<td>Calgary, AB</td>
<td>Tel: (403) 944-4057 After hours: (403) 944-1110</td>
</tr>
<tr>
<td></td>
<td>Foothills Medical Centre</td>
<td>Calgary, AB</td>
<td>Tel: (403) 944-4057 After hours: (403) 944-1110</td>
</tr>
<tr>
<td></td>
<td>Comprehensive Centre for Bleeding Disorders</td>
<td>Edmonton, AB</td>
<td>Tel: (780) 407-6588 Pager: (780) 445-1683</td>
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<tr>
<td><strong>SASKATCHEWAN</strong></td>
<td>Saskatchewan Bleeding Disorders Program</td>
<td>Saskatoon, SK</td>
<td>Tel: (306) 655-6504 After hours: (306) 655-6424 Pager - RN: (306) 655-1000 #10258</td>
</tr>
<tr>
<td></td>
<td>Royal University Hospital</td>
<td></td>
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<tr>
<td><strong>MANITOBA</strong></td>
<td>MB Bleeding Disorders Program</td>
<td>Winnipeg, MB</td>
<td>Tel: (204) 787-2465 Pager: (204) 787-2071 #3346</td>
</tr>
<tr>
<td><strong>ONTARIO</strong></td>
<td>Hemophilia Program</td>
<td>Hamilton, ON</td>
<td>Tel: (905) 521-2100 #75978 24 hour: (905) 521-2100 ext 76443</td>
</tr>
<tr>
<td></td>
<td>Hamilton Health Sciences Corporation</td>
<td>Hamilton, ON</td>
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<td></td>
<td>McMaster Division</td>
<td>Hamilton, ON</td>
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<td>Hemophilia Program</td>
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<td></td>
<td>Hamilton Health Sciences Corporation</td>
<td>Hamilton, ON</td>
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<tr>
<td><strong>QUÉBEC</strong></td>
<td>Hemophilia Clinic</td>
<td>Montréal, QC</td>
<td>Tel: (514) 412-4420 After hours: (514) 412-4400 #23333</td>
</tr>
<tr>
<td></td>
<td>Ste-Justine Hospital</td>
<td>Montréal, QC</td>
<td>Tel: (514) 345-4931 #6031 Pagers: (514) 415-5573 / 5584 After hours: (514) 345-4788</td>
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<tr>
<td></td>
<td>Regional Hemophilia Centre for Eastern Quebec</td>
<td>Montréal, QC</td>
<td>Tel: (514) 345-2360</td>
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<tr>
<td></td>
<td>Hôpital de l’ Enfant Jésus</td>
<td>Québec, QC</td>
<td>Tel: (418) 649-5624</td>
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<tr>
<td><strong>NEW BRUNSWICK</strong></td>
<td>Horizon Health Network</td>
<td>Zone 1, Moncton</td>
<td></td>
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<tr>
<td></td>
<td>Hemophilia Clinic</td>
<td>Moncton, NB</td>
<td>Tel: (506) 857-5465 / 857-5467 Emergency line: (506) 874-9561 / 1-888-475-9922</td>
</tr>
<tr>
<td></td>
<td>Inherited Bleeding Disorder Clinic</td>
<td>Saint John, NB</td>
<td>Tel: (506) 648-7286 Pager: (506) 646-3757</td>
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<tr>
<td><strong>NOVA SCOTIA</strong></td>
<td>Pediatric Bleeding Disorder Clinic</td>
<td>IWK Health Centre</td>
<td>Tel: (902) 470-8752 / 470-8819 After hour emergencies: (902) 470-8888</td>
</tr>
<tr>
<td></td>
<td>Regional Hemophilia Centre for Eastern Quebec</td>
<td>Halifax, NS</td>
<td>Tel: (902) 473-5612</td>
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<tr>
<td><strong>NEWFOUNDLAND</strong></td>
<td>Bleeding Disorders Program</td>
<td>Janeway Site</td>
<td>Eastern Health St. John’s, NL Tel: (709) 777-4388 After hours Tel: (709) 777-6300</td>
</tr>
</tbody>
</table>
**ALABAMA**

- **Children's Rehabilitation Services**  
  1610 Center St. Suite A  
  Mobile, AL 36604  
  Phone: (251) 432-4560  
  Pediatric after hours: (251) 405-5115

- **Children's Rehabilitation Services**  
  PO Drawer 2328  
  Birmingham, AL 35201-2328  
  Phone: (205) 939-5900  
  Adult after hours: (205) 934-3411  
  Pediatric after hours: (205) 939-9100

**ARKANSAS**

- **Arkansas Center for Bleeding Disorders**  
  Arkansas Children's Hospital  
  800 Marshall  
  Little Rock, AR 72202  
  Phone: (501) 364-5961  
  After hours: (501) 364-1100

**CALIFORNIA**

- **Children's Hospital Oakland**  
  Div of Hematology/Oncology  
  747 52nd Street  
  Oakland, CA 94610-4131  
  Phone: (510) 428-3286

- **Children's Hospital of Central California**  
  Hematology/Oncology  
  9300 Valley Children's Place  
  Madera, CA 93638  
  Phone: (559) 353-5460  
  Pediatric after hours: (559) 353-5460

- **Children's Hospital of Los Angeles**  
  Hem/Onc 4650 Sunset Boulevard, Box #54  
  Los Angeles, CA 9002  
  Phone: (323) 669-4141  
  After hours: (323) 660-2450

- **Children's Hospital of Orange County**  
  Dept of Hem/Oncology  
  455 South Main Street  
  Orange, CA 92868  
  Phone: (714) 699-1100  
  After hours: (714) 699-1100

- **City of Hope National Medical Center**  
  Hemophilia Trt Center  
  1500 E. Duarte Rd  
  MOB 4 Duarte, CA 91010  
  Phone: (626) 301-8426

- **Orthopaedic Hospital of Los Angeles**  
  Hemophilia Program  
  2400 S. Flower Street  
  Los Angeles, CA 90007  
  Phone: (213) 742-1357  
  After hours: (213) 742-1162

**COLORADO**

- **Mountain States Regional Hemophilia and Thrombosis Center**  
  PO Box 6507 MS F416  
  Aurora, CO 80045-0507  
  Phone: (303) 724-0362  
  Adult after hrs: (303) 372-0000  
  Ped. After hours: (303) 861-6740

**CONNECTICUT**

- **UCONN Hemophilia Treatment Center**  
  Univ of Conn Health Center  
  University Cancer Center  
  263 Farmington Ave.  
  Farmington, CT 06030  
  Phone: (860) 679-2576  
  Adult after hours: (860) 679-2000

**DELaware**

- **Christiana Care Health Services**  
  Hemophilia Program, L-214  
  Christiana Hospital  
  4755 Ogletown-Stanton Rd.  
  Newark, DE 19718  
  Phone: (302) 733-3542  
  Adult after hours: (302) 737-7700
Hemophilia Treatment Centers - USA

DISTRICT OF COLUMBIA

Children’s Healthcare of Atlanta & Scottish Rite
Aflac Cancer Ctr & Blood Disorders Service
5455 Meridian Mark Rd, Ste 400
Atlanta, GA 30342
Phone: (404) 785-3240
After hours: (404) 785-3240

Emory University Hemophilia Program Office
2015 Uppergate Dr. NE
Atlanta, GA 30322
Phone: (404) 727-1608
Adult after hours: (404) 778-5000
Pediatric after hours: (404) 778-5000

Emory University Cancer Center, Division of Hem/Onc
3800 Ponce De Leon Blvd, NW
Atlanta, GA 30308
Phone: (404) 727-1608
Adult after hours: (404) 778-5000
Pediatric after hours: (404) 778-5000

FLORIDA

All Children’s Hospital
Ped Hem/Onc Associates
880 6th St So Ste 140
St. Petersburg, FL 33701
Phone: (727) 767-4176
Pediatric after hours: (727) 527-6862

Miami Comprehensive Hemophilia Center - Pediatrics
University of Miami
Dept. of Pediatrics (R-131)
Miami, FL 33101
Phone: (305) 585-5635
Pediatric after hours: (305) 585-5400

Nemours Children’s Clinic
Division of Ped. Hem/Onc
807 Children’s Way
Jacksonville, FL 32207
Phone: (904) 390-3789
Pediatric after hours: (904) 390-3600

University of South Florida - Adult
James A. Haley V.A. Hospital
Hematology-111-R
13000 Bruce B. Downs Blvd
Tampa, FL 33612
Phone: (813) 972-7582

HAWAII

Hemophilia and Thrombosis Center of Hawaii
Kapi’olani Med Center for Women and Children
1319 Punahou Street
Honolulu, HI 96826
Phone: (808) 983-8551
After hours: (808) 524-2575

IDAHO

Idaho Regional Hemophilia Center
Mountain States Tumor Inst.
Peds. Hem/Onc
100 East Idaho Street
Boise, ID 83712-6297
Phone: (208) 381-2782
After hours: (208) 327-8007

ILLINOIS

Children’s Memorial Hospital
2300 Children’s Plaza, Box 30
Chicago, IL 60614
Phone: (773) 880-3977
Pediatric after hours: (773) 880-4000

Comprehensive Bleeding Disorders Center
5019 North Executive Drive
Peoria, IL 61614
Phone: (309) 692-4533
After hours: (309) 636-8998

John H. Stroger, Jr. Hospital of Cook County - Hemophilia Treatment Centers
1900 W. Polk St.
Chicago, IL 60612
Phone: (312) 864-4167
Adult after hours: (312) 864-1300
Pediatric after hours: (312) 864-1500

Northwestern University
Northwestern Center for Bleeding Disorders
676 N. St Clair Suite 850
Chicago, IL 60611
Phone: (312) 695-6180
Adult after hours: (312) 695-0990

Rush University Medical Center
Section of Pediatric Hematology/Oncology
Suite 718 PBI
1723 W. Harrison Street
Chicago, IL 60612-3833
Phone: (312) 942-8114
After hours: (312) 942-1674

Stroger Children’s Hospital of Cook County
Dept of Ped Hem/Oncology
700 S. Wood St., Rm. 7206
Chicago, IL 60612
Phone: (312) 864-4167
Adult after hours: (312) 864-1300
Pediatric after hours: (312) 864-1500

INDIANA

Indiana Hemophilia and Thrombosis Center
8402 Harcourt Rd, Suite 420
Indianapolis, IN 46260
Phone: (317) 871-0000 Ext. 236
Adult after hours: (877) 256-8837
Pediatric after hours: (317) 871-0000
<table>
<thead>
<tr>
<th>Hemophilia Treatment Centers - USA</th>
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<tr>
<td><strong>IOWA</strong></td>
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<tr>
<td>University of Iowa Hospitals &amp; Clinics</td>
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<tr>
<td>Iowa Reg. Hemophilia Center</td>
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<tr>
<td>Dept of Ped 2507 JCP</td>
</tr>
<tr>
<td>Iowa City, IA 52242</td>
</tr>
<tr>
<td>Phone: (319) 356-4277</td>
</tr>
<tr>
<td>After hours: (319) 356-1616</td>
</tr>
</tbody>
</table>

| **KENTUCKY**                      |
| Brown Cancer Center              |
| Hemophilia Treatment Center      |
| 529 South Jackson Rm 229         |
| Louisville, KY 40202             |
| Phone: (502) 595-4582            |
| Adult after hours: (502) 562-4053 |
| Pediatric after hours: (502) 595-4673 |

| Norton Kosair Children's Medical Center  |
| 200 E. Chestnut Street             |
| Louisville, KY 40492               |
| Phone: (502) 629-7750              |
| After hours: (502) 629-7750        |

| University of Kentucky             |
| Hemophilia Treatment Center        |
| J457 Kentucky Clinic              |
| 740 South Limestone Street         |
| Lexington, KY 40536-0284           |
| Phone: (800) 333-7359              |
| After hours: (859) 323-5321        |

| **LOUISIANA**                      |
| Louisiana Ctr for Bleeding and Clotting Disorders  |
| Tulane Univ. School of Med.        |
| 1430 Tulane Av Box TB-31           |
| New Orleans, LA 70112              |
| Phone: (504) 988-5433              |
| After hours: (504) 988-5433        |

| **MAINE**                          |
| Maine Medical Center              |
| Maine Hemophilia & Thrombosis Center  |
| 100 US Route 1, Unit 104          |
| Scarborough, ME 04074             |
| Phone: (207) 885-7683             |
| Adult after hours: (207) 885-7683  |
| Pediatric after hours: (207) 885-7565 |

| **MARYLAND**                       |
| Johns Hopkins University Medical Center  |
| 1125 Ross                           |
| 720 Rutland Avenue                 |
| Baltimore, MD 21205                |
| Phone: (304) 614-0834              |
| Adult after hours: (410) 955-6070   |
| Pediatric after hours: (410) 232-9037 |

| **MASSACHUSETTS**                  |
| Boston Hemophilia Center           |
| Brigham and Women's                |
| Brigham & Women's Hospital         |
| BWH Mid Campus- 3                  |
| 75 Francis Street                  |
| Boston, MA 02115                   |
| Phone: (617) 732-5844              |

| New England Hemophilia Center      |
| UMass Memorial Hospital           |
| 119 Belmont Street                |
| Worcester, MA 01605               |
| Phone: (800) 955-8252             |
| After hours: (800) 955-8252       |

| **MICHIGAN**                       |
| Cascade Hemophilia Consortium      |
| 210 East Huron, Suite C2           |
| Ann Arbor, MI 48104                |
| Phone: (734) 996-3300              |

| Children's Hospital of Michigan    |
| Hemostasis &Thrombosis Ctr         |
| 3901 Beaubien Blvd.                |
| Detroit, MI 48201                  |
| Phone: (313) 745-5690              |
| Pediatric after hours: (313) 745-5111 |

| DeVos Children's Hospital          |
| DeVos Childrens Coagulation        |
| Disorders Program                  |
| 100 Michigan Street, N.E., MC #85   |
| Grand Rapids, MI 49503             |
| Phone: (616) 391-2033              |
| Pediatric after hours: (616) 391-1774 |

| DMC Karmanos Cancer Institute     |
| Comprehensive Center for Bleeding Disorders and Thrombosis  |
| 4100 John R 4 Hudson Webber       |
| Detroit, MI 48201                 |
| Phone: (313) 576-8707             |
| Adult after hours: (313) 745-5111  |

| Eastern Michigan Hemophilia Treatment Center  |
| Hurley Medical Center                    |
| One Hurley Plaza                        |
| Flint, MI 48503-5993                    |
| Phone: (800) 257-9432                   |
| Adult after hours: (810) 762-8200       |
| Pediatric after hours: (810) 257-9000   |

| Hemophilia Clinic of West Michigan Cancer Center  |
| 200 North Park Street                     |
| Kalamazoo, MI 49007                       |
| Phone: (269) 373-7479                     |
| Adult after hours: (269) 373-7488         |
| Pediatric after hours: (269) 341-6350     |

| Henry Ford Hospital                   |
| Adult Hemophilia and Thrombosis Treatment Center  |
| 2799 West Grand Boulevard             |
| K-13 Hematology / Oncology            |
| Detroit, MI 48202-2689                 |
| Phone: (313) 916-3790                  |
| Adult after hours: (313) 916-2600      |

| Mich. State University                |
| Center for Bleeding Disorders and Clotting Disorders  |
| 2900 Hannah Blvd Room 202             |
| East Lansing, MI 48823                 |
| Phone: (517) 353-9385                  |

| Munson Medical Center                |
| 1105 Sixth Street                    |
| Traverse City, MI 49684              |
| Phone: (231) 935-7227                 |
| Adult after hours: (231) 935-5000     |
| Pediatric after hours: (800) 468-6766 |

| University of Michigan Hemophilia and Coagulation Disorders  |
| F2480 Mott                                    |
| 1500 East Medical Center Drive               |
| Ann Arbor, MI 48109-0235                     |
| Phone: (734) 936-6393                        |
| After hours: (734) 936-6267                  |

<p>| West Michigan Pediatric at Bronson        |
| Ped Hematology/Oncology                   |
| 601 John St, Suite E. 300                 |
| Kalamazoo, MI 49007                       |
| Phone: (269) 341-6350                     |</p>
<table>
<thead>
<tr>
<th>State</th>
<th>Center Name</th>
<th>Address</th>
<th>Phone</th>
<th>After hours</th>
<th>Pediatric after hours</th>
</tr>
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<tbody>
<tr>
<td>MINNESOTA</td>
<td>Mayo Comprehensive Hemophilia Center</td>
<td>200 First St. SW Hilton 106, Rochester, MN 55905</td>
<td>(800) 344-7726</td>
<td>(507) 284-2511</td>
<td></td>
</tr>
<tr>
<td></td>
<td>University of Minnesota Medical Center, Fairview</td>
<td>MMC 713, 420 Delaware St., SE, Minneapolis, MN 55455</td>
<td>(612) 626-6455</td>
<td>(612) 273-3000</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Hemophilia and Thrombosis Center</td>
<td>Adult after hours: (612) 273-3000, Pediatric after hours: (612) 813-5940</td>
<td></td>
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<td></td>
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<tr>
<td>MONTANA</td>
<td>Mountain States Regional Hemophilia and Thrombosis Center</td>
<td>PO Box 6507 MS F416, Aurora, CO 80045-0507</td>
<td>(303) 724-0362</td>
<td>(303) 372-0000</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Adult after hours: (303) 372-0000, Ped. After hours: (303) 861-6740</td>
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<tr>
<td>NEBRASKA</td>
<td>Nebraska Regional Hemophilia Treatment Center</td>
<td>University of Nebraska Medical Center, Omaha, NE 68198-7680</td>
<td>(402) 559-4227</td>
<td></td>
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<tr>
<td></td>
<td>Adult after hours: (732) 235-6542, Pediatric after hours: (732) 828-3000</td>
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<tr>
<td>NEVADA</td>
<td>Hemophilia Treatment Center of Las Vegas</td>
<td>Children's Center for Cancer &amp; Blood Diseases, Las Vegas, NV 89109</td>
<td>(702) 732-0971</td>
<td>(702) 732-2011</td>
<td></td>
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<tr>
<td></td>
<td>Adult after hours: (702) 732-2011, Pediatric after hours: (702) 732-0971</td>
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<tr>
<td>NEW HAMPSHIRE</td>
<td>Dartmouth-Hitchcock Hemophilia Center</td>
<td>One Medical Center Drive, Lebanon, NH 03756</td>
<td>(603) 650-5522</td>
<td>(603) 650-5000</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Adult after hours: (716) 896-2470, Pediatric after hours: (716) 343-6776</td>
<td></td>
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<tr>
<td>NEW YORK</td>
<td>Hemophilia Center of Western New York - Adult</td>
<td>Erie County Medical Center, 462 Grider Street, Buffalo, NY 14215</td>
<td>(716) 878-7446</td>
<td>(716) 896-2470</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Adult after hours: (716) 878-7446, Pediatric after hours: (716) 866-2470</td>
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<tr>
<td></td>
<td>Hemophilia Center of Western New York - Ped.</td>
<td>Children's Hospital of Buffalo, 219 Bryant St., Buffalo, NY 14222</td>
<td>(716) 878-7446</td>
<td>(716) 866-2470</td>
<td></td>
</tr>
<tr>
<td></td>
<td>Adult after hours: (716) 878-7446, Pediatric after hours: (716) 866-2470</td>
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<tr>
<td></td>
<td>Long Island Jewish Medical Center Hemophilia Treatment Center</td>
<td>Oncology Institute, Room 358, New Hyde Park, NY 11040</td>
<td>(718) 470-7380</td>
<td>(718) 343-6776</td>
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<tr>
<td></td>
<td>Adult after hours: (718) 470-7380, Pediatric after hours: (718) 343-6776</td>
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<tr>
<td>NEW MEXICO</td>
<td>Ted R. Montoya Hemophilia Center</td>
<td>Univ of N M Dept of Pediatrics, Albuquerque, NM 87121</td>
<td>(505) 272-6420</td>
<td>(505) 272-2111</td>
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<tr>
<td></td>
<td>Adult after hours: (505) 272-2111, Pediatric after hours: (505) 272-4461</td>
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<tr>
<td>MISSISSIPPI</td>
<td>University of Mississippi Medical Center</td>
<td>Ped Hematology/Oncology, 350 W Woodrow Wilson Dr, Jackson, MS 39213</td>
<td>(601) 984-2710</td>
<td>(601) 984-1000</td>
<td></td>
</tr>
<tr>
<td>MISSOURI</td>
<td>Saint Louis University Center for Bleeding and Thrombotic Disorders</td>
<td>Hemophilia Treatment Center, Adult Program</td>
<td>(314) 577-6168</td>
<td>(314) 268-5643</td>
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<tr>
<td></td>
<td>St. Louis University Hospital, West Pavilion</td>
<td>St. Louis, MO 63110</td>
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<td></td>
<td>Adult after hours: (505) 272-6420, Pediatric after hours: (505) 272-4461</td>
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<tr>
<td></td>
<td>Hemophilia Treatment Center</td>
<td>One Hospital Dr 7W12, Columbia, MO 65212</td>
<td>(573) 882-9355</td>
<td>(573) 882-4141</td>
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<tr>
<td></td>
<td>Children's Mercy Hospital</td>
<td>Adult after hours: (816) 234-3508, Pediatric after hours: (816) 404-4000</td>
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<tr>
<td></td>
<td>The John Bouhassin Center for Children with Bleeding Disorders</td>
<td>2401 Gillham Road, Kansas City, MO 64108</td>
<td>(816) 234-3508</td>
<td>(816) 404-4000</td>
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<tr>
<td></td>
<td>Adult after hours: (816) 404-4000, Pediatric after hours: (816) 234-3000</td>
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<tr>
<td></td>
<td>Mary M. Gooley Hemophilia Center, Inc.</td>
<td>1415 Portland Avenue, Suite 425, Rochester, NY 14621</td>
<td>(585) 922-5700</td>
<td>(585) 399-1717</td>
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<tr>
<td></td>
<td>Adult after hours: (585) 922-5700, Pediatric after hours: (585) 399-1717</td>
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</table>
Hemophilia Treatment Centers - USA

Mount Sinai School of Medicine Regional Comprehensive Hemophilia Treatment Center
19 East 98th Street, Suite 9D
Box 1078 New York, NY 10029
Phone: (212) 876-8701
After hours: (212) 876-8701

SUNY Upstate Medical University
Adult Program
c/o Regional Onc Center
750 E. Adams Street
Syracuse, NY 13210
Phone: (315) 464-8200
Adult after hours: (315) 464-8200

SUNY Upstate Medical University
Pediatric Program
c/o Center for Children with Cancer & Blood Disorder
750 E. Adams St, Rm 5400
Syracuse, NY 13210
Phone: (315) 464-5294
Pediatric after hours: (315) 701-1790

The Regional Comp. Hemophilia & von Willebrand Trmt Ctr.
at Albany Medical College
47 New Scotland Ave A 52
Albany, NY 12208
Phone: (518) 626-5827 or 800-773-7080
Adult after hours: (518) 786-7723
Pediatric after hours: (518) 262-5513

UHSH Blood Disorder Center
UHS Wilson Hospital
33-57 Harrison Street
Johnson City, NY 13790
Phone: (607) 763-6436
After hours: (607) 763-6000

Well Medical College of Cornell University Regional Comprehensive Hemophilia Diagnostic and Treatment Center
525 E. 68th St, Room P-695
New York, NY 10021
Phone: (212) 746-3418
Adult after hours: (212) 746-2927
Pediatric after hours: (212) 746-3400

East Carolina University
Brody School of Medicine
PCMH 288 West
Greenville, NC 27858-4354
Phone: (252) 744-4676

Univ. of N. Carolina at Chapel Hill School of Medicine
W1022 Old Clinic Building
CB # 7016
Chapel Hill, NC 27599-7016
Phone: (919) 966-4736
After hours: (919) 966-4736

Wake Forest University
School of Medicine
Baptist Medical Center
The Bowman Gray Campus
Department of Pediatrics
Medical Center Boulevard
Winston-Salem, NC 27157-1081
Phone: (336) 716-4324
Adult after hours: (336) 713-5440
Pediatric after hours: (336) 716-4324

North Dakota

MeritCare Hospital
DBA Roger Maris Cancer Center
Hemophilia & Thrombosis Treatment Center
820 Fourth Street North
Fargo, ND 58122
Phone: (701) 234-7544
Pediatric after hours: (701) 234-6000

Ohio

Children's Hospital
Medical Center of Akron
Hemophilia Treatment Center
One Perkins Square
Akron, OH 44308-1062
Phone: (330) 543-8732

Cincinnati Children's Hospital
Medical Center
Hemophilia Treatment Center
3333 Burnet Avenue
Cincinnati, OH 45229
Phone: (513) 636-4269
Pediatric after hours: (513) 636-4200

Columbus Children's Hospital
Columbus Hemophilia Treatment Center
700 Children's Drive
Columbus, OH 43205
Phone: (614) 722-3240
Pediatric after hours: (614) 722-3250

Dayton Children's Medical Center
West Central Ohio Hemophilia Treatment Center
One Children's Plaza
Dayton, OH 45404-1815
Phone: (937) 641-5877
Adult after hours: (937) 641-3000
Pediatric after hours: (937) 641-3000

Oregon Hemophilia Treatment Center
OR Health & Science Univ.
707 SW Gaines Rd
Portland, OR 97239-2901
Phone: (503) 494-8716
After hours: (503) 494-9000

Northwest Ohio Hemophilia Treatment Center
The Toledo Hospital
Children's Medical Center
2150 W. Central Avenue
Toledo, OH 43606
Phone: (419) 291-2210
Adult after hours: (419) 473-3200
Pediatric after hours: (419) 291-8520

Ohio State University Medical Center Hemophilia Treatment Center
M414 Starling Loving Hall
320 W. 10th Avenue
Columbus, OH 43210
Phone: (614) 293-8183
Adult after hours: (614) 293-8000

Mount Sinai School of Medicine Regional Comprehensive Hemophilia Diagnostic and Treatment Center
19 East 98th Street, Suite 9D
Box 1078 New York, NY 10029
Phone: (212) 876-8701
After hours: (212) 876-8701

SUNY Upstate Medical University
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Phone: (315) 464-8200
Adult after hours: (315) 464-8200

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Pediatric Program
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Pediatric after hours: (518) 262-5513

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UHS Wilson Hospital
33-57 Harrison Street
Johnson City, NY 13790
Phone: (607) 763-6436
After hours: (607) 763-6000

Well Medical College of Cornell University Regional Comprehensive Hemophilia Diagnostic and Treatment Center
525 E. 68th St, Room P-695
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Pediatric after hours: (212) 746-3400

Ohio State University Medical Center Hemophilia Treatment Center
M414 Starling Loving Hall
320 W. 10th Avenue
Columbus, OH 43210
Phone: (614) 293-8183
Adult after hours: (614) 293-8000

UHHS Cleveland
Univ Hospitals Health System
Ped Hem Mail Stop 6054
1100 Euclid Avenue
Cleveland, OH 44106
Phone: (216) 844-3345
Adult after hours: (216) 844-8220
Pediatric after hours: (216) 844-3345

University of Cincinnati Medical Center
Hemophilia Treatment Center
231 Albert Sabin Way
Mail Location 562
Cincinnati, OH 45267-0562
Phone: (513) 584-7639
Adult after hours: (513) 584-7661

Oklahoma

Oklahoma Center for Bleeding Disorders
940 N.E. 13th St, Rm. 3B3308
Oklahoma City, OK 73104
Phone: (405) 271-3661
Adult after hours: (405) 271-4222
Pediatric after hours: (405) 271-5437

Oregon Hemophilia Treatment Center
OR Health & Science Univ.
707 SW Gaines Rd
Portland, OR 97239-2901
Phone: (503) 494-8716
After hours: (503) 494-9000
### PENNSYLVANIA

<table>
<thead>
<tr>
<th>Center Name</th>
<th>Address</th>
<th>Phone Numbers</th>
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<tbody>
<tr>
<td>Cardeza Foundation Hemophilia Center</td>
<td>Thomas Jefferson University Hospital</td>
<td>111 S. 11th Street, Philadelphia, PA 19107; Phone: (215) 955-8435, Adult after hours: (215) 955-8874</td>
</tr>
<tr>
<td>Children's Hospital of Philadelphia</td>
<td>Hemophilia Program</td>
<td>34th St. &amp; Civic Center Blvd 4th Floor Wood Building, Philadelphia, PA 19104; Phone: (215) 590-4493, Pediatric after hours: (215) 590-1000</td>
</tr>
<tr>
<td>Hemophilia Center of Central Pennsylvania</td>
<td>The Milton S. Hershey Med. Center</td>
<td>500 University Drive, PO Box 850, H046, Hershey, PA 17033; Phone: (717) 531-7468, Adult after hours: (717) 531-8521</td>
</tr>
<tr>
<td>Lehigh Valley Hospital</td>
<td>Hemophilia Treatment Center</td>
<td>1240 South Cedar Crest Blvd. Suite 103, Allentown, PA 18105-1556; Phone: (610) 402-0640, Adult after hours: (610) 402-7880</td>
</tr>
<tr>
<td>Penn Comprehensive Hemophilia Program</td>
<td>Univ. of Pennsylvania Med. Ctr - Presbyterian</td>
<td>51 N. 39th St. MAB Ste 106, Philadelphia, PA 19104; Phone: (215) 662-9960, Adult after hours: (215) 662-4000</td>
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### SOUTH CAROLINA

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<tr>
<td>Palmetto Health Richland</td>
<td>Hemophilia Center of South Carolina</td>
<td>7 Richland Medical Park Rd. Suite 203, Columbia, SC 29201-6872; Phone: (803) 434-3533, After hours: (803) 434-3533</td>
</tr>
<tr>
<td>South Dakota Center for Blood Disorders</td>
<td>Sioux Valley Children's Specialty Clinic</td>
<td>1305 West 18th Street, Sioux Falls, SD 57117-5039; Phone: (605) 333-7171, After hours: (605) 333-7188</td>
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### SOUTH DAKOTA

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<tr>
<th>Center Name</th>
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<tbody>
<tr>
<td>Galveston Hemophilia Program</td>
<td>University of TX Med. Branch</td>
<td>Rm 4.160 John Sealy Annex, Galveston, TX 77555-0565; Phone: (409) 772-1165</td>
</tr>
<tr>
<td>Gulf States Hemophilia and Thrombophilia Center</td>
<td></td>
<td>6655 Travis, Suite 400, Houston, TX 77030; Phone: (713) 500-8360, After hours: (713) 704-4284</td>
</tr>
<tr>
<td>North Texas Comprehensive Hemophilia Center</td>
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<td>Adult Program</td>
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<td>North Texas Comprehensive Hemophilia Center-Pediatric Program</td>
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<td>South Texas Hemophilia &amp; Thrombophilia Trt. Center</td>
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<td>Texas Children's Hemophilia and Thrombophilia Center</td>
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### TENNESSEE

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<tr>
<td>East Tennessee Comprehensive Hemophilia Center</td>
<td>University of Tennessee Medical Center</td>
<td>4 North West, 1924 Alcoa Highway, Knoxville, TN 37920-6999; Phone: (865) 544-9170, After hours: (865) 544-9170</td>
</tr>
<tr>
<td>St. Jude Research Hospital</td>
<td></td>
<td>332 N. Lauderdale, Memphis, TN 38101-0318; Phone: (901) 448-6454, Adult after hours: (901) 448-6454, Pediatric after hours: (901) 448-6454</td>
</tr>
<tr>
<td>University of Tennessee - Memphis Hemophilia Clinic of Memphis University of Tennessee</td>
<td></td>
<td>920 Madison Ave Suite 300, Memphis, TN 38103-3446; Phone: (901) 448-6454, After hours: (901) 448-6454</td>
</tr>
<tr>
<td>Vanderbilt University Medical Center</td>
<td>Hemostasis-Thrombosis Clinic</td>
<td>2220 Pierce Ave 397 PRB, Nashville, TN 37232-6310; Phone: (615) 936-1765, Adult after hours: (615) 936-1803, Pediatric after hours: (615) 936-1765</td>
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### UTAH

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<th>Center Name</th>
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<tr>
<td>Mountain States Hemophilia &amp; Thrombophilia Center</td>
<td></td>
<td>100 N. Medical Drive, Salt Lake City, UT 84113; Phone: (801) 588-3477, Adult after hours: (801) 581-2121, Pediatric after hours: (801) 588-2000</td>
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### SOUTH CAROLINA

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<tr>
<td>Galveston Hemophilia Program</td>
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<td>University of TX Med. Branch</td>
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<td>Gulf States Hemophilia and Thrombophilia Center</td>
<td></td>
<td>6655 Travis, Suite 400, Houston, TX 77030; Phone: (713) 500-8360, After hours: (713) 704-4284, Or (713) 536-1152</td>
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<tr>
<td>North Texas Comprehensive Hemophilia Center</td>
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<td>Adult Program</td>
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<tr>
<td>North Texas Comprehensive Hemophilia Center-Pediatric Program</td>
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<tr>
<td>South Texas Hemophilia &amp; Thrombophilia Center</td>
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<tr>
<td>Texas Children's Hemophilia and Thrombophilia Center</td>
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<tr>
<td>Vanderbilt University Medical Center</td>
<td>Hemostasis-Thrombosis Clinic</td>
<td>2220 Pierce Ave 397 PRB, Nashville, TN 37232-6310; Phone: (615) 936-1765, Adult after hours: (615) 936-1803, Pediatric after hours: (615) 936-1765</td>
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### TEXAS

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<th>Center Name</th>
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<tbody>
<tr>
<td>Fort Worth Bleeding Disorders Program</td>
<td>Cook Children's Medical Center</td>
<td>901 Seventh Avenue, Suite 220, Ft. Worth, TX 76104; Phone: (682) 885-4007, Pediatric after hours: (682) 885-4000</td>
</tr>
</tbody>
</table>
Hemophilia Treatment Centers - USA

VERMONT

Vermont Regional Hemophilia Center
108 Cherry St PO Box 70
Burlington, VT 05402
Phone: (802) 865-1326

WASHINGTON

Puget Sound Blood Center & Program
Hemophilia Program
921 Terry Avenue
Seattle, WA 98104-1256
Phone: (206) 292-6507
After hours: (206) 292-6525

WEST VIRGINIA

Charleston Area Medical Center
c/o Cancer Care Center
3200 MacConkle Avenue, SE
Charleston, WV 25304
Adult after hours: (877) 541-9446

WISCONSIN

Comprehensive Center for Bleeding Disorders
The Blood Center of Southeastern Wisconsin
PO Box 2178
Milwaukee, WI 53201-2178
Phone: (414) 257-2424
After hours: (414) 257-2424

WYOMING

Mountain States Regional Hemophilia and Thrombosis Center
PO Box 6307 MS F416
Aurora, CO 80045-0507
Phone: (303) 724-0362
Adult after hrs: (303) 372-0000
Ped. After hours: (303) 861-6740

U.S. TERRITORIES

Guam Comprehensive Hemophilia Care Program
Department of Public Health & Social Services
PO Box 2816
Hagatna, GU 96932
Phone: (671) 735-7168

Puerto Rico
Hemophilia Treatment Center
University of Puerto Rico
School of Medicine
Department of Pediatrics Box 5067
San Juan, PR 00936
Phone: (787) 777-3535 Ext. 7013/4

Internet Resources

Canadian Hemophilia Society
www.hemophilia.ca

National Hemophilia Foundation
1-800-42-HANDI
www.hemophilia.org

World Federation of Hemophilia
www.vhf.org

CDC
www.cdc.gov

Emergency Care for Patients with Hemophilia
www.HemophiliaEmergencyCare.com

Project Red Flag
www.ProjectRedFlag.org
Emergency Care for Patients with von Willebrand Disease

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**Introduction & von Willebrand Disease Basics**

**Purpose**

This manual contributes to von Willebrand Disease (VWD) care by enhancing the emergency department personnel’s understanding of this disorder and its treatment. The goals of this manual are to:

- promote understanding of the complexities of von Willebrand Disease with an emphasis on rapid treatment for correction of the hemostatic abnormality
- provide a reference for the emergency center staff
- promote a consultative dialogue with the emergency department (ED), treatment center, and patient/family

**Early triage and treatment reduce morbidity.**

**Use**

This manual provides a standardized format for evaluation and treatment of VWD emergencies. The content is segmented by systems and complications of VWD. Turn to an area of interest. The illustration on the left page provides information points for quick review. The text on the right page gives further detail of bleeding presentations, their possible complications and treatment. The treatment varies to the type and the severity of VWD. Treatment and management information is provided on the inside cover of the manual as a reference.

It is suggested that the patient’s treatment center or hematologist be consulted for final management of bleeding complications.

**To The Attending Medical Staff**

This manual is a guide for medical personnel who may be less familiar with VWD treatment. The content consists of guidelines, recommendations and suggestions only. The attending physician has the final responsibility for appropriate diagnosis and treatment.

**Definition**

von Willebrand Disease is an autosomally-inherited bleeding disorder caused by the quantitative deficiency or dysfunction of von Willebrand factor, a large multimeric glycoprotein. It is non-sex linked. Therefore, it can occur equally in both men and women.

**Effects of von Willebrand Disease**

von Willebrand factor is essential for platelet-plug formation as an adhesion protein that diverts circulating platelets to the sites of vascular injury, particularly through larger multimers. It also forms a non-covalent complex with coagulation factor VIII in plasma, thereby protecting it from inactivation and clearance.

Even though the primary deficiency or defect in von Willebrand Disease is that of von Willebrand factor, the secondary deficiency of factor FVIII, which is dependent on von Willebrand factor as its naturally occurring plasma carrier and stabilizer, leads to a defect both in platelet-plug formation and in fibrin formation.

**Prevalence**

The prevalence is as high as 1 to 2 percent in the general population.
Introduction & von Willebrand Disease Basics

Types of von Willebrand Disease

The type of VWD determines the treatment - see inside of the front cover for treatment options. von Willebrand Disease is classified by ‘type 1, 2 or 3’. If the type is unknown, proceed as if type 1. If bleeding continues contact a hematologist.

von Willebrand Disease is classified into three main phenotypes and each have subtypes based on the quantity and quality of the von Willebrand factor (VWF):

- **Type 1**: which accounts for 60 to 80 percent of cases, results from a decreased production of normal von Willebrand factor and factor VIII; typically transmitted as an autosomal dominant trait in the heterozygous state.

- **Type 2**: which accounts for 10 to 30 percent of cases is characterized by qualitative abnormalities of von Willebrand factor and is further divided into subtypes 2A, 2B, 2M and 2N. Inheritance is generally autosomal dominant.

- **Type 3**: Accounts for 1 to 5 percent of cases and is transmitted as an autosomal recessive trait in homozygous or compound heterozygous persons. This severe form of the disease is characterized by a very low or undetectable von Willebrand factor in plasma with a low, usually detectable factor VIII activity. **It is in these rare cases of type 3 (1 in 1 million people) that symptoms are more frequent and severe, similar to those cases of severe hemophilia.**

Acquired von Willebrand Disease: This is an acquired syndrome that resembles von Willebrand Disease in its clinical manifestation and laboratory patterns. It occurs in rare instances in association with clinical conditions such as lymphoproliferative and autoimmune diseases, hypothyroidism, essential thrombocytopenia, cancer, Wilm’s tumor and valvular heart disease.

Bleeding episodes

The hallmark of von Willebrand Disease is mucosal bleeding. Mucous membrane bleeds such as bleeding from the nose, mouth, gastrointestinal tract, genitourinary and vaginal bleeding are the most common. If left untreated, these mucous membrane bleeds can become acute and sometimes life-threatening emergencies. Serious bleeding resulting from untreated trauma and/or post-surgical bleeding can also become life or limb-threatening in these patients.

Serious bleeding sites

The major sites of serious bleeding which threaten life, limb, or function are:

- intracranial
- oropharynx
- vaginal bleeding
- ocular
- spinal cord
- gastrointestinal
- intra-abdominal

Treatment

The mainstay of treatment is the replacement of the deficient/defective protein at the time of bleeding or before invasive procedures are performed. This may require desmopressin (subcutaneous, intranasal, or intravenous) or an infusion of commercial von Willebrand factor/FVIII concentrate such as Humate-P®, Alphanate® and wilate®. Specific doses, additional drugs and medical interventions depend upon the type of VWD and the site and severity of bleeding. Please refer to the inside cover of the manual for more detailed information on the recommended treatment. Once treatment has been given, emergency diagnostic procedures can begin.

Family

Patients living with VWD or their parents are often knowledgeable about the management of their disorder and their input should be sought and heeded. Interview the family about whether any medication has been administered prior to arriving at the ED; if so, determine when and what dose. Additional treatment may be required, dependent on the time lag and severity of the bleed. Determine the treating hematologist or treatment center, and contact them for assistance and follow-up as needed.
Intracranial hemorrhage (ICH) is a potential for all head injuries.

Administer recommended treatment first*, and then perform diagnostic studies such as CT scan.

If an ICH is diagnosed, the patient should be admitted and the hematologist contacted immediately. If no ICH is diagnosed, the patient may be discharged.

Discharge Instructions

Call the treatment center or the patient’s hematologist for follow-up treatment recommendations*.

Report any signs or symptoms of an ICH to the treatment center or the patient’s hematologist.

Head injury instructions should be given for a two week period (instead of the usual instructions for 24 - 48 hour period).
Intracranial hemorrhage (ICH) is a potential risk for individuals with von Willebrand Disease, and is most commonly associated with injury. The risk of intracranial hemorrhage is increased with the more severe types of VWD. Without early recognition and treatment, death or severe neurologic impairment can occur. Early neurologic symptoms may not always be evident.

**Treatment**

All significant head trauma, with or without hematoma, should be treated promptly with the appropriate treatment before any diagnostic tests. A hematologist should be contacted.

**Diagnostic imaging**

Obtain an emergency CT scan to rule out ICH after the appropriate treatment has been given. Notify the patient’s hematologist or treatment center as soon as possible.

**Possible admission**

The patient should be admitted to the hospital for observation if he/she has suffered a severe blow to the head or exhibits any neurologic symptoms. Symptoms can include headache with increasing severity, irritability, vomiting, seizures, vision problems, focal neurologic deficits, stiff neck, or changes in level of consciousness. **Patients with a past history of ICH are at increased risk of repeated head bleeds.**

**Instructions**

If the patient is discharged home, instruct the family to monitor the patient for signs and symptoms of neurologic deterioration and report any abnormalities to the hematologist. Consult the treatment center to arrange for follow-up treatment if the patient is discharged home from the emergency department.
Mucous Membrane Bleeding

A Dental or E.N.T. consult may be needed.

Nose bleeds may respond to other measures. Refer to “Controlling Epistaxis” table on pg. 8.

Mouth bleeds (gum, tooth, frenulum or tongue laceration) may require treatment* and the use of anti-fibrinolytics. If the bleeding is minor, local measures such as topical thrombin (if available) or fibrin glue (Tisseal®) in conjunction with anti-fibrinolytics can be used. Refer to the anti-fibrinolytics on pg. 9.

Assess for anemia if there has been prolonged mucosal bleeding.

Discharge Instructions

Patients should follow-up with their treatment center or hematologist the next day.

Instruct the patient on how to control epistaxis, the use of anti-fibrinolytics, and the importance of a modified diet. Consult the Diet Modifications table on pg. 9 as needed.
Mucous membrane bleeding may require medical care in the emergency department. Treatment may be required for patients who:

- are experiencing profuse and/or prolonged bleeding
- have sustained a known injury to the mouth, tongue, or nose
- have severe swelling in the mouth or throat area
- are experiencing respiratory distress
- have difficulty swallowing or speaking

The patient may not know the reason for the symptom or bleeding. It may have been caused by trauma, infection, or the bleed may be spontaneous. If airway blockage is suspected, prompt treatment is required prior to any invasive procedures.

**Remember prompt treatment will greatly reduce the bleeding, often preventing serious complications.** The longer the patient waits, the more bleeding takes place. If the bleed is in a closed space, the accumulation of blood will cause surrounding tissue damage, airway obstruction and pain.

**Epistaxis**

Uncontrolled epistaxis may require treatment in conjunction with anti-fibrolytics. Be sure the patient knows how to control and stop the bleeding (see pg. 8)

**Oral Cavity**

Bleeding in the mouth can be hard to control. A frenulum or tongue laceration may respond to topical thrombin or other similar agents. If the bleed continues, the patient will probably need further treatment. A single treatment may temporarily stop the bleeding, but clot lysis from saliva enzymes often results in re-bleeding. Re-bleeding is most commonly seen on days 3-5. An anti-fibrinolytic may be indicated to maintain hemostasis. A modified diet should be started at the same time as treatment (see Diet Modifications pg. 9).

Bleeding may occur with extracted, erupting or exfoliating teeth. It is more common with extracted and exfoliating teeth. A dental consult may be needed to extract the tooth. Treatment to increase the von Willebrand factor will be necessary prior to extraction. A frenulum or tongue laceration will require treatment.

**Retropharyngeal**

After the recommended treatment, further observation, X-rays and admission may be required depending upon the specific circumstance.
Mucous Membrane Bleeding

Controlling Epistaxis
Instruct the patient:

1. To gently blow his/her nose to remove mucus and unstable clots that will interfere with hemostasis.

2. Tilt the head forward so any blood will come out the nares and not down the back of the throat.

3. Apply firm constant pressure to the entire side of the nose that is bleeding for 15 minutes.

4. Release the pressure to see if bleeding has stopped, gently blow out and remove any soft clots.

5. If the bleeding continues, reapply pressure for another five minutes.

6. Recommended treatment* and/or anti-fibrinolytic agents (see next page) may be needed.

7. During active bleeding, NoseBleed QR® powder is an over-the-counter preparation can be utilized. The powder needs to be mixed with blood, as per the manufacturer's directions. The powder will solidify the blood, form a crust and bleeding may stop.

8. During active bleeding, or when the bleeding has stopped, you may spray or apply two drops of oxymetazoline (eg. NeoSynephrine®, Dristan® or Afrin®) nasal spray/drops to the side that was bleeding. These can be used at home PRN for epistaxis.

9. Instruct the patient to use mucosal membrane moisturizer (eg. Vaseline®, Secaris®) in the nares to keep the membranes soft and moist, and prevent the formation of hard crusts which might crack and restart bleeding. Adequate humidification in the home is also helpful.

10. An Ear Nose Throat (ENT) consult may be required for possible cauterization of a vessel.
Anti-Fibrinolytics

Anti-fibrinolytics may be indicated in nasal or oral bleeding. Amicar® and Cyklokapron® are both anti-fibrinolytic agents. Either may be prescribed for mucous membrane bleeding to promote clot stabilization in conjunction with the recommended treatment*. In some cases they may be prescribed independently.

Amicar - epsilon aminocaproic acid  
Recommended dosage:
Child: oral dose 50-100 mg/kg (not to exceed 4 g) every 6 hours for 3 - 10 days
Adult: oral dose 3-4 g every 6 hours for 3 -10 days
Supplied: Tablet: 500 mg or 1000 mg per tab
Syrup: 250 mg per ml
Injectable: 250 mg /ml available in 20 ml vial
*Contraindicated if hematuria present

Cyklokapron - tranexamic acid  
Recommended dosage:
Child: oral dose 25 mg/kg every 6- 8 hours for 3 - 10 days
Adult dose: oral dose 1000 mg-1500 mg  tid for 3 - 10 days
Supplied: Tablet: 500 mg tranexamic acid per tab
Injectable: 100 mg/ml available in 5 and 10 ml ampules
*Contraindicated if hematuria present

These medications must be given as ordered to keep blood levels constant. They are not readily available through local pharmacies (they must be ordered). If possible, dispense the amount for 2-3 days from the hospital pharmacy to allow time for the local pharmacy to order. Other options are the family’s home supply, bleeding disorders treatment center or (U.S.) home care companies.

Follow-up care per the treatment center or patient’s hematologist. Topical agents such as topical Thrombin® and Gelfoam® may also be used to help control mucous membrane bleeding.

Diet Modifications
Directions for the patient:

1. Diet should be restricted to soft, cool, or lukewarm foods until the area is fully healed. Suggested foods: flavored gelatin, non-carbonated drinks, sherbet, lukewarm soups (no cream soups), baby foods, blenderized or pureed foods, pasta.

2. Avoid using a straw, chewing gum, and do not smoke. Negative pressure from the sucking action can dislodge the clot and aggravate the bleeding site.

3. Foods to avoid include hard foods like chips, nuts, popcorn, tacos, etc.

4. If Desmopressin (DDAVP®, Octostim®, or Stimate®) has been utilized for treatment, the patient has fluid restrictions for 24 hours.
Nausea and vomiting may indicate intracranial hemorrhage as well as gastrointestinal problems.

**Iliopsoas bleeding**
- flexed hip
- pain on extension
- Management: Treatment product* as per hematologist

**Abdominal pain**
Treat immediately* as per hematologist for:
- flank pain
- melena
- vomiting blood

**Hematuria**
- bed rest for 24 hours
- force fluids
- consult the treatment center or the patient’s hematologist
- avoid anti-fibrinolytics

**Discharge Instructions**
- increase fluids
- rest
- no heavy lifting
- report any symptoms such as fever, pain, or increased hematuria, melena, hematemesis
- follow-up with the treatment center or the patient’s hematologist

*Recommended treatment table inside front cover
Initial presentation

Acute abdominal pain in a patient with von Willebrand Disease may have many origins, such as gastrointestinal (GI) tract hematomas (both spontaneous or trauma induced), iliopsoas or retroperitoneal bleeding.

Bleeding may also occur with hemorrhoids or the passage of kidney stones. Notify the treatment center or the patient’s hematologist.

Patients who present to the emergency department with abdominal or flank pain, melena or hematemesis should be triaged for immediate examination and the recommended treatment should be initiated. Once this is done, then diagnostic x-rays, scans and endoscopy procedures can be carried out.

Abdominal trauma and benign events such as forceful coughing or vomiting can precipitate an abdominal bleed. Blood loss can be significant before outward signs and symptoms appear. Infants can have bleeds with gastroenteritis, intussusception or Meckel's Diverticulum.

A history of lifting heavy objects, weight lifting, falling on a bicycle crossbar or stretching the groin can precipitate abdominal wall, iliopsoas (see pg. 14 and 15), or retroperitoneal bleeding. These types of bleeds can occur more commonly in individuals with type 3 VWD, and are rarely seen in type 1 and type 2 VWD.

Symptoms

Symptoms of abdominal muscle bleeding (rectus, pectorals, latissimus, obliques) are a palpable mass, rigidity, and pain. Concurrent bleeding in the abdominal cavity may be present and go unnoticed for days with a steadily dropping hemoglobin. Rupture of the liver, spleen, or pancreas should be considered when the hemoglobin falls dramatically following trauma.

For nausea and vomiting without an obvious cause, consider that these may be symptoms of intracranial bleeding. Inquire about head injury, mental status changes, and other neurologic signs and symptoms, and consider CT scan of the head.

Genitourinary bleeding

Hematuria is often frightening to the patient but not a serious event. Instruct the patient to remain at bed rest and to increase fluids to 16 oz or 500 ml every hour over the next 24 hours. Protracted hematuria may require treatment.

Anti-fibrinolytics are contraindicated with hematuria. Contact the hematologist.

Scrotal bleeding may occur after trauma, especially in toddlers. Treatment will be required and follow-up with the hematologist or treatment center should be arranged.
Gynecological Bleeding

Assess for signs of anemia
- check hemoglobin
- check ferritin level

Obtain accurate menstrual history
- pad and/or tampon count per hour or per 24 hr time period (include nights)
- frequency of changing protection
- amount of blood on each pad (use a pictorial chart if available)
- presence of clots, size of clots
- number of overflow or flooded pads
- length, regularity of menses
- missed days at school/work due to menses
- need for iron therapy either currently or in the past

For active menorrhagia:
In addition to Humate-P®, Alphanate and wilate® or Desmopressin (DDAVP® or Stimate®/Octostim® as preparation available), start an anti-fibrinolytic (pg. 9).

Consider prescribing birth control therapy or IV Premarin as adjunctive therapy to prevent more bleeding.

Discharge Instructions
- follow-up with the treatment center or the patient’s hematologist (within one week)
- Instruct patient to accurately record bleeding, menstrual history
- Recommend rest, drinking fluids, eating iron rich foods (or use supplemental iron preparations)
Menstrual Bleeding

Prolonged and heavy menstrual bleeding is one of the most common symptoms for females with bleeding disorders.

**Menarche** – a teenage girl with von Willebrand Disease can present to an emergency department at menarche or soon after with a severe, occasionally life-threatening hemorrhage. Appropriate treatment should commence immediately. Major vaginal bleeding requires treatment with a von Willebrand factor concentrate (ex. Humate-P®*, Alphanate® and wilate®) Consultation with an OB-GYN specialist and hematologist at a bleeding disorder treatment center is essential for ongoing follow-up. Oral contraceptives, anti-fibrinolytic treatment and desmopressin (IV, intranasally or subcutaneously) may be recommended on an ongoing basis.

Assess for signs of anemia, as the patient's hemoglobin can drop 2-3 g/ml Hgb, in just a few days, from prolonged menses.

**Menses** - Some women bleed excessively through their menstrual cycle. Others bleed between cycles or continuously through the month. These women may present to the ED with menorrhagia, iron deficiency, anemia, or mittelschmerz due to increased bleeding with ovulation. Obtain an accurate menstrual history and contact the patient’s hematologist for treatment recommendations.

Assess for signs of anemia, as the patient's hemoglobin can drop to 2-3 g/ml Hgb, in just a few days, from prolonged menses.

**Postpartum Bleeding** – During pregnancy, the majority of women with von Willebrand Disease, type 1, will have normal von Willebrand factor and factor VIII levels due to increased estrogen levels. “There are very few published data on the use of desmopressin during pregnancy, but there are some concerns that desmopressin causes uterine contraction with premature labour, intrauterine growth retardation and hyponatremia. For these reasons, it is advisable to be cautious about the use of desmopressin during pregnancy. Once the cord is clamped, desmopressin can be used if necessary. It is also probably reasonable to use desmopressin before a caesarean. Desmopressin is not contraindicated during lactation.”


The von Willebrand factor levels will decrease 24 to 48 hours following delivery, thereby increasing the risk of post-partum bleeding. In the event of a post-partum hemorrhage, treatment should be initiated immediately to elevate the von Willebrand factor levels. Life-threatening post-partum hemorrhage will require treatment with a von Willebrand factor/FVIII concentrate (ex. Humate-P®*, Alphanate® and wilate®) Adjunctive treatment with intravenous or oral anti-fibrinolytics may be useful (see pg. 9).
**Soft Tissue / Muscle / Joint Bleeding**

**Neck swelling:** EMERGENCY
- potential airway compromise
Management: Treatment product* as per hematologist

**Soft tissue bleeds and bruising**
- no functional impairment
- tenderness, but no severe pain
Management: No treatment, R.I.C.E.*

**Iliopsoas bleeds**
- flexed hip
- pain / inability to extend the leg on the affected side
Management: Treatment product* as per hematologist

**Deltoid / forearm bleed**
- increased swelling and bruising
- observe for symptoms of compartment syndrome
Management: Treatment product* as per hematologist

**Thigh/calf/buttock bleed**
- pain
- with/without swelling
- impaired mobility
- observe for signs and symptoms of compartment syndrome
Management: Treatment product* as per hematologist

**Early onset joint bleed**
- tingling - pain
- limited range of motion

**Advanced joint bleed**
- heat - pain
- swelling
Management: Treatment product* as per hematologist. Ice and immobilization for comfort.

**Thigh/calf/buttock bleed**
- pain
- with/without swelling
- impaired mobility
- observe for signs and symptoms of compartment syndrome
Management: Treatment product* as per hematologist

**Discharge Instructions**

- "RICE - Rest, Ice, Compression (Ace® wraps), Elevation
- Crutches - for weight bearing joints and crutch instructions
- Sling or splinting if support is needed (i.e. Aircast® for ankles)
- follow-up with the treatment center or the patient’s hematologist

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*Recommended treatment table inside front cover
Soft tissue and superficial bleeds

Soft tissue bleeds usually do not require aggressive treatment. Superficial hematomas and bruises respond well to rest, ice and elevation. If the hematoma and bruising continue to increase in size, impairing movement or function, treatment may be required.

Muscle bleeds

Muscle bleeding is usually only associated with trauma in persons with mild von Willebrand Disease.

Persons with the most severe type of von Willebrand Disease, type 3, can experience muscle bleeding spontaneously or with minimal trauma. Any muscle group may be subject to bleeding. Common bleeding sites include the upper arm, forearm, thigh, and calf muscles.

Muscles that exhibit warmth, pain, and swelling should be managed with the recommended treatment. Anti-fibrinolytics may also be helpful.

Consequences of muscle bleeds: Muscle bleeds can result in serious consequences if not treated promptly. Extensive blood loss may occur in large muscle groups. Muscle bleeding can place pressure on nerves and blood vessels and, if untreated, may result in permanent disabilities such as foot drop and wrist contracture. It is important that the patient’s hematologist be consulted before any invasive procedures.

Treatment and follow-up care: Occasionally muscle bleeds may require treatment but more often will resolve with conservative treatment such as rest and ice. If compartment syndrome is suspected, appropriate treatment should be initiated and the patient should be admitted with an emergency consult to hematology.

Joint Bleeding

Joint bleeding is uncommon in individuals with type 1 and 2 von Willebrand Disease and is usually associated with trauma. Individuals with type 3 von Willebrand Disease can experience joint bleeding with or without trauma, and bleeding can occur into any joint space.

The joints most commonly affected are the elbows, knees, and ankles. Less common sites include the shoulders and hips. As repeated bleeding occurs, the synovial tissue thickens and develops even more friable blood vessels. A vicious cycle of bleeding and rebleeding may set in and the affected joint is referred to as a “target joint.” Eventually, repeated bleeding into joints leads to arthropathy with destruction of cartilage and the eventual erosion of bone. The end result is decreased joint mobility and function.

Signs and symptoms: Outward signs of joint bleeding include restriction of movement, swelling, heat, and erythema on and around the joint. The patient may report symptoms of a bubbling or tingling sensation with no physical signs. Later symptoms include a feeling of fullness within the joint and moderate to severe pain as the bleed worsens.

Treatment: Some patients may present for treatment with no other outward signs of bleeding than decreased range of motion and a complaint of pain or tingling. This is indicative of an early onset joint bleed and is the optimal time to treat. The patient should be infused as quickly as possible with the recommended treatment in order to minimize pain and joint destruction. Extreme pain, swelling, heat, and immobility are signs and symptoms of an advanced joint bleed which occurs only after blood has filled the joint space.

Initiate treatment before any diagnostic procedures such as x-ray. Before dislocated joints are reduced, infuse with the recommended treatment.

Joint Aspiration: Caution!

The aspiration of joint bleeds in VWD is contraindicated unless recommended by the treatment center.
Desmopressin is a synthetic form of antidiuretic hormone which causes the release of factor VIII and von Willebrand factor from the endothelial cell storage sites. It can increase the VWF level by as much as three to five fold.

Desmopressin is the preferred treatment for type 1 VWD and certain patients with type 2. The response to desmopressin can vary greatly with each individual. Therefore, prior to use, a desmopressin trial should be done with results reviewed and recorded by a hematologist. If the patient does respond to desmopressin, the full effect is reached 30 to 90 minutes after administration and hemostasis is maintained for approximately 24 hours. If the patient does not respond to desmopressin, hemostasis can be maintained with infusions of factor concentrates containing both von Willebrand protein and factor VIII, such as Humate – P®, Alphanate®, and wilate®.

Expected side effects: short term facial flushing, increased heart rate, red conjunctiva, and headache.

Dose

SC/IV: 0.3 mcg/kg/dose. Recommendation: a maximum dose of 20 mcg.

IV ROUTE: Dilute with normal saline (50-100 ml). Infuse over 30-60 minutes. No less than 30 minutes. It is recommended to administer the medication with the individual in a supine position.

SC ROUTE: The subcutaneous route is advantageous to minimize drug side effects.

Supplied

Ampules: 4 mcg/ml DDAVP®, 15 mcg/ml OCTOSTIM®

Nasal Spray

OCTOSTIM® or STIMATE® nasal spray must be brand specific to ensure patient receives the correct dose of DDAVP® that will stop the bleeding.

150 mcg / 0.1 ml per single spray (OCTOSTIM®, STIMATE®)

Recommended dose for patients over 50 kg: 300 mcg (1 spray per nostril, total of two sprays)

Recommended dose for patients under 50 kg: 150 mcg (1 spray in one nostril, total of one spray)

Unreliable absorption if the intranasal route is compromised. Hematologist should be consulted for treatment guidelines.

Indications

Treatment of von Willebrand Disease Type 1 and certain forms of Type 2

Contraindications

Hypersensitivity, infants under 3 months, patients suffering from dehydration, history of seizure, coronary artery insufficiency.

Recommended treatment table inside front cover
Use With Caution

- Elderly
- Patients with von Willebrand Disease Type 2B
- Young children especially under 2 years of age
- Hypertensive cardiovascular disease
- Individuals with low-normal blood pressure

Desmopressin has an antidiuretic effect. **Patients should be advised to avoid alcohol and restrict their fluid intake to thirst only for 24 hours after receiving the drug.** Infants and children will require careful fluid intake restriction to prevent possible hyponatremia and water intoxication. Accurate intake and output should be recorded on any patients receiving IV fluids.

Adverse Effects

**Cardiovascular:** facial flushing, sweating, dizziness, transient hypertension, hypotension, and tachycardia, hyponatremia.

**Gastrointestinal:** nausea, vomiting.

**Neurologic:** headache, tremor, seizures.

**Local:** pain and erythema at injection site or in nasal mucosa if intranasal spray is used.

**Thrombocytopenia:** in Type 2B von Willebrand Disease

### Topical Preparations

<table>
<thead>
<tr>
<th>Amicar 10% topical solution</th>
<th>Mix 2 ml Amicar (IV preparation 250 mg/ml), and 3 ml sterile water for injection</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Soak gauze in solution, squeeze out excess and apply to area.</td>
</tr>
<tr>
<td></td>
<td>Discard solution after 24 hours</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Tranexamic Acid 5% topical solution</th>
<th>Mix 5 ml Tranexamic acid, use the IV preparation 100mg/ml (5 ml ampule size) and 5 ml sterile water for injection</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Soak gauze in solution, squeeze out excess and apply to area.</td>
</tr>
<tr>
<td></td>
<td>Discard solution after 24 hours</td>
</tr>
</tbody>
</table>

**Request from Pharmacy**

Tranexamic Acid 5% Nasal Gel: To make a 5% Tranexamic Acid Nasal gel, take ten (10) Tranexamic acid 500 mg tablets and crush with a very small amount of 70% alcohol. Measure out 100 grams of Intrasite gel (methylcellulose). Gradually add the Intrasite gel to the crushed tablets/paste. Once mixed put in an ointment jar. Stable for 10 days refrigerated (probably much longer). Apply with Q-tip® or finger once or twice a day.

Topical agents such as topical Thrombin® and Gelfoam® and fibrin glue (Tisseal-R) may also be used to help control mucous membrane bleeding.

Antibiotics and pain medications may also be indicated in the treatment of mucosal bleeds.
**Factor Administration**

Reconstitute per package insert.

Products containing von Willebrand factor are plasma derived products.

The volume may be 10 ml, 20 ml, 30 ml

Examples: Humate-P®, Alphanate®, wilate®

Labeled: Antihemophilic Factor/von Willebrand Factor Complex (Human) Dried, Pasteurized

**Mixing instructions and the rate of administration are found on the drug insert.**

It is best to follow treatment recommendations that the patient may carry or to consult with the patient’s treatment center.

**Dosage**

Each bottle of factor concentrate is labeled with the activity expressed in both von Willebrand ristocetin co-factor international units (vWF:RCo I.U.) and factor VIII international units (F VIII I.U.).

The dosage to be administered is based on the patient’s body weight in kilograms (kg) and is normally ordered in ristocetin co-factor units (VWF:RCo I.U.). The von Willebrand factor/FVIII concentrate is a plasma-derived factor that has been virally inactivated.

The ENTIRE contents of all the vials reconstituted for an infusion should be used, even if it exceeds the calculated dosage. A larger dose will only prolong the period of normal coagulation. Due to its cost, factor concentrate should never be discarded!

Document the lot number(s), expiration date(s), factor concentrate trade name and total number of units infused. This information can be found on the factor concentrate's box.

Some patients are instructed to bring unmixed factor concentrate with them to the ED to minimize treatment delay and cost. Occasionally, patients will bring prepared factor concentrate after unsuccessful home venipuncture attempts. Please assist with venipuncture and allow the patient or family to infuse the prepared factor concentrate, if possible, per your institution’s policy.
Other Medications

Routine medications

Patients with VWD can receive routine medications (e.g. pain medications, antibiotics, etc.) that do not interfere with clotting function. Avoid non-steroidal anti-inflammatories (NSAIDS), ASA and any product with aspirin-related ingredients (e.g. Pepto-Bismol®, Excedrin®, Percodan®).

Medications for fever or pain

Acetaminophen can be given for fever or pain. Narcotics/opioids can be given to control pain experienced by the patient with a bleeding disorder. Avoid giving intramuscular injections of pain medications because of the possibility of causing a muscle bleed.

Routes of administration

Medications which can be given PO, SC, or IV are preferred. If the rabies vaccination series is needed, an experienced hematologist (preferably the patient’s) should be contacted for advice prior to and after the injections in order to prevent internal bleeding.

For any needle stick, pressure for a minimum of 5 minutes afterward will minimize soft tissue or muscle bleeding. Avoid giving intramuscular injections of antibiotics, pain medications, or immunizations because of the possibility of causing a muscle bleed. You can also apply an ice pack for 15 - 20 minutes.

Caution

Some patients with VWD may have liver disease from hepatitis or may have been exposed to HIV. Use caution when prescribing drugs that may cause liver toxicity or could cause potential serious drug interactions.
Treatment should never be delayed for laboratory studies to be drawn or completed.

**Head injury**

First give the recommended treatment...  
...then perform a CT scan.

**Fracture**

First give the recommended treatment...  
...then immobilize appropriately.

**Discharge Instructions**

Patient should follow-up with the treatment center or hematologist the next day.  
Head injury: Discharge with routine post head injury instructions (patient should be assessed for two weeks instead of 48 hours).
In general, patients with VWD who are experiencing an acute bleeding episode may need treatment as well as basic first aid measures. Do not delay treatment to perform testing.

**Laboratory studies**

If the only complaint is an acute joint or muscle bleed, no laboratory studies are necessary. If GI, uterine, or oral cavity bleeding is suspected and has potentially been extensive, a complete blood count may be indicated to determine if the individual is anemic. Treatment should never be delayed for laboratory studies to be drawn or completed.

**X-rays and other radiological studies**

Remember that a swollen joint or extremity can be the result of internal bleeding. X-rays of the joint can be used to document a joint bleed, but are generally not useful in detecting early onset bleeds when treatment is optimal.

A CT of the head (see pg. 4) is necessary when dealing with a potential intracranial hemorrhage. Give the maximum recommended treatment before sending the patient to CT scan.

**Fractures**

Give the recommended treatment, then X-ray and set the bone.

**Lacerations and sutures**

Sutures and staples can be used. If the laceration is significant enough to require sutures, the patient should first receive the recommended treatment and then proceed with the procedure. Contact the patient’s hematologist for follow-up treatment instructions. No treatment is usually needed for suture removal.

**Invasive procedures**

Invasive procedures should be performed as clinically indicated, i.e. lumbar puncture with symptoms of meningitis. However, factor replacement treatment should be given prior to the procedure.

**Arterial sticks and venipunctures**

Do not perform arterial sticks unless no other option is available. If an arterial stick must be done, then the recommended treatment and precautions should be taken before the procedure begins.

Venipuncture may be done at any location; hands are generally excellent and no pre-treatment is necessary. Avoid “digging” for deep veins. Apply pressure for several minutes or until there is no further oozing noted at the venipuncture and IV removal sites.
Many different emergencies / trauma may occur to persons with von Willebrand Disease, just as to others.

- Animal bites
- Burns
- Falls
- Fractures (see pg. 20)

- Motor vehicle accidents
- Gunshot wounds
- Ocular injuries
- Puncture wounds

**Treatment**

For any serious injury, a major dose of a factor VIII product containing von Willebrand factor (eg. Humate-P®, Alphanate®, Wille®) should be infused prior to blood work, CT scan, X-rays or other scans, debriding, sutures, etc.

For less serious injuries, other treatment options may suffice and can be considered: local treatment (pg. 8), desmopressin (pg. 16-17), anti-fibrinolitics (pg. 9).


Kasper, Carol K. (2004). Von Willebrand Disease.[Monograph]. Los Angeles, USA:.Orthopedic Hospital, [Published by Aventis Behring Foundation for research and advancement of patient care].


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