Emergency Care for Patients with von Willebrand Disease

An instructional manual for Medical Professionals

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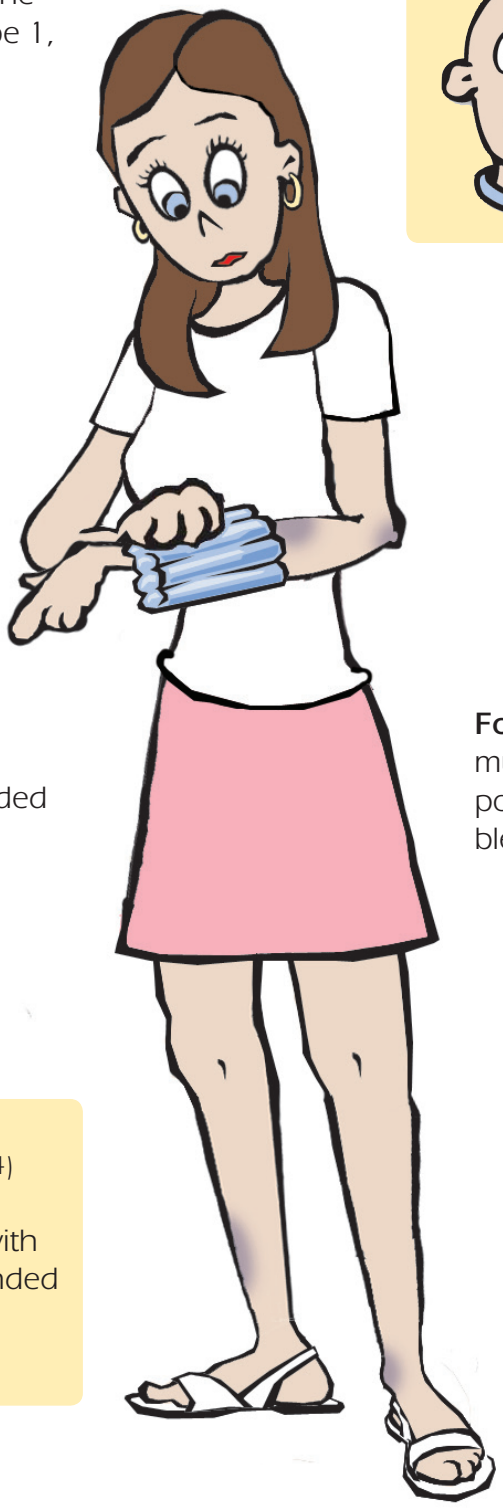
von Willebrand Disease (VWD) is classified by 'type 1, 2, or 3' If the type is unknown proceed as if type 1, if bleeding continues consult a hematologist.

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

Gynecological bleeds (pg. 12)



Immobilizers
p.r.n. for joint bleeds

Ice pack
for soft tissue, muscle, 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

Treatment and Management Guidelines for von Willebrand Disease

Type of von Willebrand Disease	Major life-threatening bleeds (ex. - head injury, GI bleeding, severe menorrhagia, etc.)	Other bleeds (ex. - sutures, nosebleed, mouth bleed, dental extractions etc.)
Type 1 or Type 2 The type 2 VWD known as 'pseudo VWD or VWF platelet type' will only respond to a platelet transfusion - call a hematologist.	Factor concentrate containing both FVIII (8) and von Willebrand factor (eg. Humate P®): 60-80 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.	**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®): 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.
Type 3 Most severe form of VWD.	Factor concentrate containing both FVIII (8) and von Willebrand factor (eg. Humate P®): 60-80 Ristocetin cofactor units/kg 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.	Factor concentrate containing both FVIII (8) and von Willebrand factor (eg. Humate P®): 40-60 Ristocetin cofactor units/kg 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.

If your institution does not have Humate-P®, but does have Alphanate® or 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.

Hemophilia Treatment Centres - Canada

BRITISH COLUMBIA

Hemophilia Program of BC (Adult Division)
St. Paul's Hospital
Vancouver, BC
Tel: (604) 682-2344, ext. 63026
After hours: (604) 682-2344

Pediatric Hemophilia/ Hematology
BC Children's Hospital
Vancouver, BC
Tel: (604) 875-2345 ext. 5335
Pager: (604) 875-2161
After hours: (604) 875-2161

ALBERTA

Southern Alberta Hemophilia Program
Alberta Children's Hospital
Calgary, AB
Tel: (403) 955-7311
After hours: (403) 955-7070

Comprehensive Centre for Bleeding Disorders
University of Alberta Hospital/
Stollery Children's Hospital
Edmonton, AB
Tel: (780) 407-6588
Pager: (780) 445-1683

SASKATCHEWAN

Saskatchewan Bleeding Disorders Program
Royal University Hospital
Saskatoon, SK
Tel: (306) 655-6504
After hours: (306) 655-1000

MANITOBA

MB Bleeding Disorders Program
Health Sciences Centre
Winnipeg, MB
Tel: (204) 787-2465
Pager: (204) 787-2071 #3346

ONTARIO

Hemophilia Program
Hamilton Health Sciences Corporation
McMaster Division
Hamilton, ON
Tel: (905) 521-2100 #75978
24 hour: (905) 521-2100 ext 76443

Bleeding Disorders Program
London Health Science Ctr.
Victoria Hospital
London, ON
Tel: (519) 685-8500 ext. 53582

Hemophilia Program
Thunder Bay Regional Hospital
Science Centre
Thunder Bay
Tel: (807) 684 6550

Comprehensive Hemophilia
Care Centre
St. Michael's Hospital
Toronto, ON
Tel: (416) 864-5129
Pager: (416) 685-9404/9478
After hours: (416) 864-5431

Hemophilia Program
Hospital for Sick Children
Toronto, ON
Tel: (416) 813-5871
Pager: (416) 377-9716
After hours: (416) 813-7500

Hematology Clinic
Children's Hospital of Eastern Ontario
Ottawa, ON
Tel: (613) 737-7600 ext. 2368

Regional Comprehensive Care Centre
for Hemophilia & Hemostasis
(Adult Program)
Ottawa Hosp. Gen. Campus
Ottawa, ON
Tel: (613) 737-8252
After hours: (613) 722-7000

Hemophilia Program, Sudbury &
North-Eastern Ontario
Laurentian Site of HRSRH
Sudbury, ON
Tel: (705) 522-2200 ext. 3264

Southeastern Ontario Regional
Inherited Bleeding Disorders Program
Kingston General Hospital
Douglas 3
Kingston, ON
Tel: (613) 549-6666 ext. 4683
Tel: 24 HR # (613) 548-7850

QUÉBEC

Hemophilia Clinic
CHUS - Hôpital Fleurimont
Sherbrooke, QC
Tel: (819) 346-1110 ext. 14560

Hemophilia Clinic
Montreal Children's Hospital
Montréal, QC
Tel: (514) 412-4420

Hemophilia Clinic - 1er vidéotron
Ste-Justine Hospital
Montréal, QC
Tel: (514) 345-4931 #6031
Pagers: (514) 415-5573 / 415-5584 /
415-5807
After hours: (514) 345-4788

Quebec Reference Centre for the
Study of Patients with Inhibitors
Hem. Clinic - 1er vidéotron
Ste-Justine Hospital
Montréal, QC
Tel: (514) 345-2360

Regional Hemophilia Centre for
Eastern Quebec
Hôpital de l' Enfant Jésus
Québec, QC
Tel: (418) 649-5624

NEW BRUNSWICK

South East Regional Health Authority
Hemophilia Clinic
Moncton, NB
Tel: (506) 857-5465 / 857-5467
Pager: (506) 558-7158

Inherited Bleeding Disorder Clinic
Saint John Regional Hospital
Saint John, NB
Tel: (506) 648-7286

NOVA SCOTIA

Pediatric Bleeding Disorder Clinic
IWK Health Centre
Halifax, NS
Tel: (902) 470-8752 / 470-8819
Pager: (902) 470-8888 #1982
After hours: (902) 470-8394

Hereditary Bleeding Disorders
Program - Adult
QE II Health Science Centre
Halifax, NS
Tel: (902) 473-5612

NEWFOUNDLAND

Hemophilia Program
Eastern Health Corporation -
Health Sciences Centre, Janeway Site
St. John's, NL
Tel: (709) 777-4388
After hours Tel: (709) 777-6300

Hemophilia Treatment Centers - USA

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

Children's Rehab. Services
407 Governor's Dr SW, Ste B
Huntsville, AL 35801
Phone: (256) 518-8650

University of Alabama
Birmingham Medical Center
1600 7th Ave S ACC 512
Birmingham, AL 35233
Phone: (205) 939-9285
Adult after hours: (205) 934-3411
Pediatric after hours: (205) 939-9100

ALASKA

Alaska Hemophilia Association
and Treatment Center
16958 N. Eagle River Loop
Eagle River, AK 99577
Phone: (907) 622-4045
After hours: (907) 268-1190

ARIZONA

Mountain States Regional
Hemophilia Center – Tucson
Univ. of AZ H/S Center
1501 North Campbell Avenue
Tucson, AZ 85724
Phone: (520) 626-7584
After hours: (520) 694-6000

Phoenix Children's Hospital
Hemophilia Ctr
O/P Building B
1919 E. Thomas Rd
Phoenix, AZ 85016
Phone: (602) 546-0920
After hours: (602) 546-0920

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) 532-8459
Adult after hours: (714) 765-6677
Pediatric after hours: (714) 765-6677

Children's Hospital, San Diego
3020 Children's Way
San Diego, CA 92123
Phone: (858) 966-5811

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

Stanford University Medical Center
Div of Hematology/Oncology
1000 Welch Road, Suite 300
Mail Code 5798
Palo Alto, CA 94304
Phone: (650) 497-8953

University of California at Davis
Hemophilia Program
2360 Stockton Blvd. Ste 1100
Davis One Building
Sacramento, CA 95817
Phone: (916) 734-3461
Adult after hours: (916) 734-2011
Pediatric after hours: (916) 734-3591

University of California, San Diego
200 W. Arbor Drive M/S 0821
San Diego, CA 92103
Phone: (619) 471-0335
Adult after hours: (619) 290-5539

University of California, San Francisco
Hemophilia Program
650 Moffitt, Box 0106
San Francisco, CA 94143
Phone: (415) 476-1280
Adult after hours: (415) 353-2421

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

Yale University School of Medicine
Yale-New Haven Hemophilia Ctr.
Dept. of Ped LMP 4087
333 Cedar Street
New Haven, CT 06510
Phone: (203) 785-4640

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 National Medical Center
Dept of Hem/Oncology
111 Michigan Avenue, NW
Washington, DC 20010
Phone: (202) 884-3622
Pediatric after hours: (202) 884-5000

Georgetown University Hospital
Lombardi Cancer Center,
Division of Hem/Onc
3800 Reservoir Road, NW
Washington, DC 20007
Phone: (202) 687-0117
Adult after hours: (202) 687-7243

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) 562-6862

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

GEORGIA

Backus Children's Hospital
4700 Waters Avenue
P.O. Box 23089
Savannah, GA 31403-3089
Phone: (912) 350-7285
Pediatric after hours: (912) 658-3017

Children's Healthcare of Atlanta at 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

Hemophilia of Georgia, Inc.
8800 Roswell Road, Ste 170
Atlanta, GA 30350
Phone: (770) 518-8272

Medical College of Georgia - Adult
Dept of Adult Hem/Onc
1120 15th St BAA-5407
Augusta, GA 30912-3125
Phone: (706) 721-0870
After hours: (706) 721-2505

Medical College of Georgia
Pediatric Hem. Program
Dept of Pediatric Hem/Onc
1446 Harper Street, BG-2013
Augusta, GA 30912-3730
Phone: (706) 721-3626

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
1725 W. Harrison Street
Chicago, IL 60612-3833
Phone: (312) 942-8114
After hours: (800) 847-1674

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

Hemophilia Treatment Centers - USA

IOWA

University of Iowa Hospitals & Clinics
Iowa Reg. Hemophilia Center
Dept of Ped 2507 JCP
Iowa City, IA 52242
Phone: (319) 356-4277
After hours: (319) 356-1616

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

Boston Hemophilia Center
Children's Hospital
Fegan 717.2
Boston, MA 02115
Phone: (617) 355-8246
Adult after hours: (617) 732-5656
Pediatric after hours: (617) 355-6101

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

West Michigan Pediatric at Bronson
Ped Hematology/Oncology
601 John St, Suite E. 300
Kalamazoo, MI 49007
Phone: (269) 341-6350

Hemophilia Treatment Centers - USA

MINNESOTA

Mayo Comprehensive
Hemophilia Center
200 First St. SW Hilton 106
Rochester, MN 55905
Phone: (800) 344-7726
After hours: (507) 284-2511

University of Minnesota
Medical Center, Fairview
Hemophilia and Thrombosis Center
MMC 713
420 Delaware St., SE
Minneapolis, MN 55455
Phone: (612) 626-6455
Adult after hours: (612) 273-3000
Pediatric after hours: (612) 813-5940

MISSISSIPPI

University of Mississippi
Medical Center
Ped Hematology/Oncology
350 W Woodrow Wilson Dr
Jackson, MS 39213
Phone: (601) 984-2710
After hours: (601) 984-1000

MISSOURI

Saint Louis University Center for
Bleeding and Thrombotic Disorders
Hemophilia Treatment Center, Adult Program
St. Louis University Hospital, West Pavilion
3655 Vista Avenue, 3rd Floor, Hem/Onc
St. Louis, MO 63110
Office: (314) 577-6168
Fax: (314) 268-5643

Hemophilia Treatment Center
One Hospital Dr 7W12
Columbia, MO 65212
Phone: (573) 882-9355
After hours: (573) 882-4141

Kansas City Regional Hemophilia Center
The Children's Mercy Hospital
2401 Gillham Road
Kansas City, MO 64108
Phone: (816) 234-3508
Adult after hours: (816) 404-4000
Pediatric after hours: (816) 234-3000

The John Bouhasin Center for
Children with Bleeding Disorders
Cardinal Glennon Children's Hospital
Saint Louis Univ. Dept of Pediatrics
1465 South Grand Blvd.
St. Louis, MO 63104
Phone: (314) 577-5332
Pediatric after hours: (314) 577-5600

MONTANA

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

NEBRASKA

Nebraska Regional
Hemophilia Treatment Center
University of Nebraska Medical Center
987680 Nebraska Medical Center
Omaha, NE 68198-7680
Phone: (402) 559-4227

NEVADA

Hemophilia Treatment
Center of Las Vegas
Children's Center for
Cancer & Blood Diseases
3059 S. Maryland Pkwy #202
Las Vegas, NV 89109
Phone: (702) 732-0971
Adult after hours: (702) 732-2011
Pediatric after hours: (702) 732-0971

NEW HAMPSHIRE

Dartmouth-Hitchcock
Hemophilia Center
One Medical Center Drive
Lebanon, NH 03756
Phone: (603) 650-5522
After hours: (603) 650-5000

NEW JERSEY

Children's Hospital of Philadelphia
Speciality Center
New Jersey Section of Hem/Onc
1012 Laurel Oak Road, Building 1014
Voorhees, NJ 08043
Phone: (856) 435-7502
Pediatric after hours: (215) 590-1000

Newark Beth Israel Medical Center
Comprehensive Hemophilia
Treatment Center
201 Lyons Avenue at Osborne Terrace
Newark, NJ 07112
Phone: (973) 926-6511
Adult after hours: (973) 926-7230
Pediatric after hours: (973) 926-7161

St. Michael's Medical Center
Nadeene Brunini Comp.
Hemophilia Care Center
111 Central Ave.
Newark, NJ 07102
Phone: (973) 877-5340
After hours: (973) 877-5340

UMDNJ-Robert Wood Johnson
Medical School
New Jersey Regional
Hemophilia Program
One Robert Wood Johnson Place,
Room #378C, CN-19
New Brunswick, NJ 08903
Phone: (732) 235-6542
After hrs: (732) 828-3000
Ask for Hem. Fellow on call

NEW MEXICO

Ted R. Montoya Hemophilia Center
Univ of N M Dept. of Pediatrics
MSC10 5590
1 University of New Mexico
Albuquerque, NM 87121
Phone: (505) 272-6420
Adult after hours: (505) 272-2111
Pediatric after hours: (505) 272-4461

NEW YORK

Hemophilia Center of
Western New York - Adult
Erie County Medical Center
462 Grider Street
1st floor, Suite 20
Buffalo, NY 14215
Phone: (716) 896-2470

Hemophilia Center of
Western New York - Ped.
Children's Hospital of Buffalo
219 Bryant St.
Buffalo, NY 14222
Phone: (716) 878-7446
Pediatric after hours: (716) 896-2470

Long Island Jewish Medical Center
Hemophilia Treatment Center
Oncology Institute, Room 358
270-05 76th Avenue
New Hyde Park, NY 11040
Phone: (718) 470-7380
After hours: (718) 343-6776

Mary M. Gooley
Hemophilia Center, Inc.
1415 Portland Avenue, Suite 425
Rochester, NY 14621
Phone: (585) 922-5700
After hours: (585) 399-1717

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) 262-5827 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

Weill 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

NORTH CAROLINA

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

Forum Health
Youngstown Hemophilia Center
Regional Referral Center
500 Gypsy Lane, 1st Floor
Youngstown, OH 44501
Phone: (330) 884-4176

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

UHHS Cleveland
Univ Hospitals Health System
Ped Hem Mail Stop 6054
11100 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

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

Hemophilia Treatment Centers - USA

PENNSYLVANIA

Cardeza Foundation Hemophilia Center
Thomas Jefferson University Hospital
Gibbon Building, Suite 4225
111 S. 11th Street
Philadelphia, PA 19107
Phone: (215) 955-8435
Adult after hours: (215) 955-8874

Children's Hospital of Philadelphia
Hemophilia Program
34th St. & Civic Center Blvd
4th Floor Wood Building
Philadelphia, PA 19104
Phone: (215) 590-4493
Pediatric after hours: (215) 590-1000

Hemophilia Center
of Central Pennsylvania
The Milton S. Hershey Med. Center
500 University Drive, PO Box 850, H046
Hershey, PA 17033
Phone: (717) 531-7468
Adult after hours: (717) 531-8521

Hemophilia Center
of Western Pennsylvania
3636 Boulevard of the Allies
Pittsburgh, PA 15213
Phone: (412) 209-7280
Adult after hours: (412) 209-7040

Lehigh Valley Hospital
Hemophilia Treatment Center
1240 South Cedar Crest Blvd. Suite 103
Allentown, PA 18105-1556
Phone: (610) 402-0640
Adult after hours: (610) 402-7880

Penn Comprehensive Hemophilia Program
Univ. of Pennsylvania
Med. Ctr - Presbyterian
51 N. 39th St. MAB Ste 106
Philadelphia, PA 19104
Phone: (215) 662-9960
Adult after hours: (215) 662-4000

RHODE ISLAND

Rhode Island Hospital
Hemophilia Center of Rhode Island
George Clinic
Providence, RI 02903
Phone: (401) 444-8250
After hours: (401) 350-9707

SOUTH CAROLINA

Palmetto Health Richland
Hemophilia Center of South Carolina
7 Richland Medical Park Rd.
Suite 203
Columbia, SC 29203-6872
Phone: (803) 434-3533
After hours: (803) 434-3533

SOUTH DAKOTA

South Dakota Center
for Blood Disorders
Sioux Valley Children's
Specialty Clinic
1305 West 18th Street
Sioux Falls, SD 57117-5039
Phone: (605) 333-7171
After hours: (605) 333-7188

TENNESSEE

East Tennessee Comprehensive
Hemophilia Center
University of Tennessee Medical Center
4 North West
1924 Alcoa Highway
Knoxville, TN 37920-6999
Phone: (865) 544-9170
After hours: (865) 544-9170

St. Jude Research Hospital
332 N. Lauderdale
Memphis, TN 38101-0318
Phone: (901) 448-6454
Adult after hours: (901) 448-6454
Pediatric after hours: (901) 448-6454

University of Tennessee - Memphis
Hemophilia Clinic of Memphis
University of Tennessee
920 Madison Ave Suite 300
Memphis, TN 38103-3446
Phone: (901) 448-6454
After hours: (901) 448-6454

Vanderbilt University Medical Center
Hemostasis-Thrombosis Clinic
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

TEXAS

Fort Worth Bleeding Disorders Program
Cook Children's Medical Center
901 Seventh Avenue, Suite 220
Ft. Worth, TX 76104
Phone: (682) 885-4007
Pediatric after hours: (682) 885-4000

Galveston Hemophilia Program
University of TX Med. Branch
Adult Hematology/Oncology
Rm 4.160 John Sealy Annex
Galveston, TX 77555-0565
Phone: (409) 772-1165

Gulf States Hemophilia and
Thrombophilia Center
6655 Travis, Suite 400
Houston, TX 77030
Phone: (713) 500-8360
After hours: (713) 704-4284
Or (713) 536-1152

North Texas Comprehensive
Hemophilia Center
Adult Program
Univ of TX Southwestern Medical School
5323 Harry Hines Blvd.
Room NC8.126
Dallas, TX 75390-8852
Phone: (214) 648-1939
After hours-Parkland: (214) 590-8000
After hours-UT Southwest: (214) 648-7070

North Texas Comprehensive
Hemophilia Center-Pediatric Program
Children's Medical Center
1935 Motor Street
Dallas, TX 75235
Phone: (214) 456-2379
Pediatric after hours: (214) 456-7000

South Texas Hemophilia &
Thrombophilia Trt. Center
Christus Santa Rosa Children's Hospital
333 N. Santa Rosa, 8th Floor
San Antonio, TX 78207
Phone: (210) 704-2187

Texas Children's Hemophilia
and Thrombophilia Center
6701 Fannin Ste. 1420
Houston, TX 77030
Phone: (832) 822-4242
After hours: (832) 824-2099

UTAH

Mountain States
Regional Hemophilia Center
Primary Children's Medical Center
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

Hemophilia Treatment Centers - USA

VERMONT

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

VIRGINIA

Children's Hospital of the King's Daughters Bleeding Disorders Center of Hampton Roads
601 Children's Lane
Norfolk, VA 23507
Phone: (757) 668-7243
Pediatric after hours: (757) 668-7243

University of Virginia Hospital
Box 800386, Pediatric Hematology
Charlottesville, VA 22908
Phone: (434) 924-8499
Adult after hours: (434) 924-0000
Pediatric after hours: (434) 924-0211

University of Virginia Hospital Adult Hemophilia Program
Hem/Onc Box 800747
Jordan Hall, Room 2353
Charlottesville, VA 22908
Phone: (434) 243-5809
Adult after hours: (434) 924-0000
Pediatric after hours: (434) 924-0211

Virginia Commonwealth University West Hospital
1200 E. Broad Street
4th Fl, Rm 442, Southwing
Richmond, VA 23298-0461
Phone: (804) 827-3306
Adult after hours: (804) 828-0951

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

West Virginia University Medical Center Robert C. Byrd Health Sciences Center Mary Babb Randolph Cancer Center
PO Box 9162
Morgantown, WV 26506
Adult after hours: (877) 427-2894

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

Gundersen Clinic
1836 South Avenue
LaCrosse, WI 54601
Phone: (608) 782-7300
Adult after hours: (800) 362-9567
Pediatric after hours: (800) 362-7567

Hemophilia Outreach Centre
1794 E. Allouez Ave.
Green Bay, WI 54311
Phone: (920) 965-0606
After hours: (920) 965-0606

UWHC Comprehensive Program for Bleeding Disorders
2704 Marshall Court
Madison, WI 53705
Phone: (608) 890-9495
After hours: (608) 262-0486

WYOMING

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

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.wfh.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

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.

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 thrombocythemia, 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
- spinal cord
- oropharynx
- gastrointestinal
- vaginal bleeding
- intra-abdominal
- ocular

Treatment

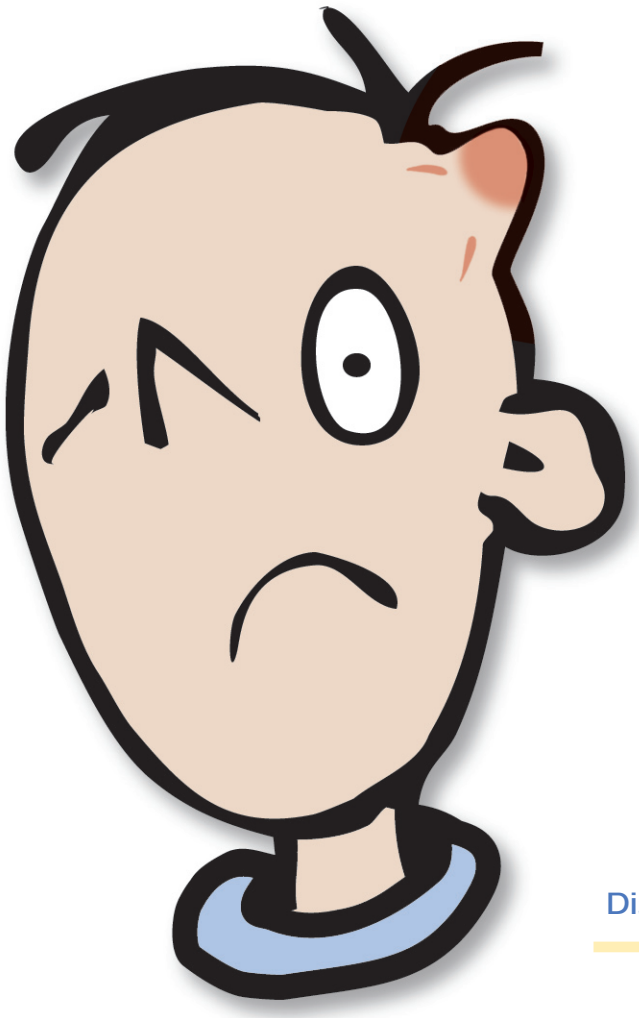
The mainstay of treatment is the replacement of the deficient/defective protein at the time of bleeding or before invasive procedures are performed. This may require desmopressin (subcutaneous, intranasal, or intravenous) or an infusion of commercial von Willebrand factor/FVIII concentrate such as Humate-P®. 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.

Head Injury

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).

 *Recommended treatment table inside front cover

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.

 *Recommended treatment table inside front cover

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

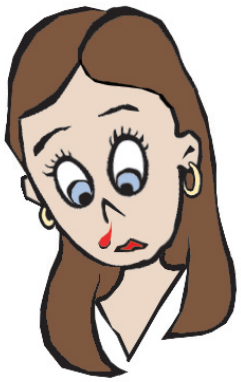
Bleeding in the mouth can be hard to control. A frenulum or tongue laceration may respond to topical thrombin or other similar agents. If the bleed continues, the patient will probably need further treatment. A single treatment may temporarily stop the bleeding, but clot lysis from saliva enzymes often 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  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:

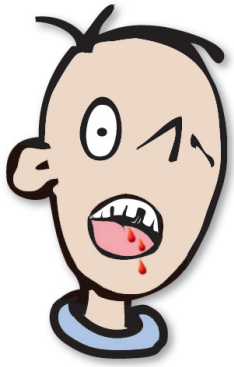
Child: oral dose 25 mg/kg every 6- 8 hours for 3 - 10 days
Adult dose: oral dose 1000 mg-1500 mg tid for 3 - 10 days
Supplied: Tablet: 500 mg tranexamic acid per tab
Injectable: 100 mg/ml available in 5 and 10 ml ampules
***Contraindicated if hematuria present**

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.

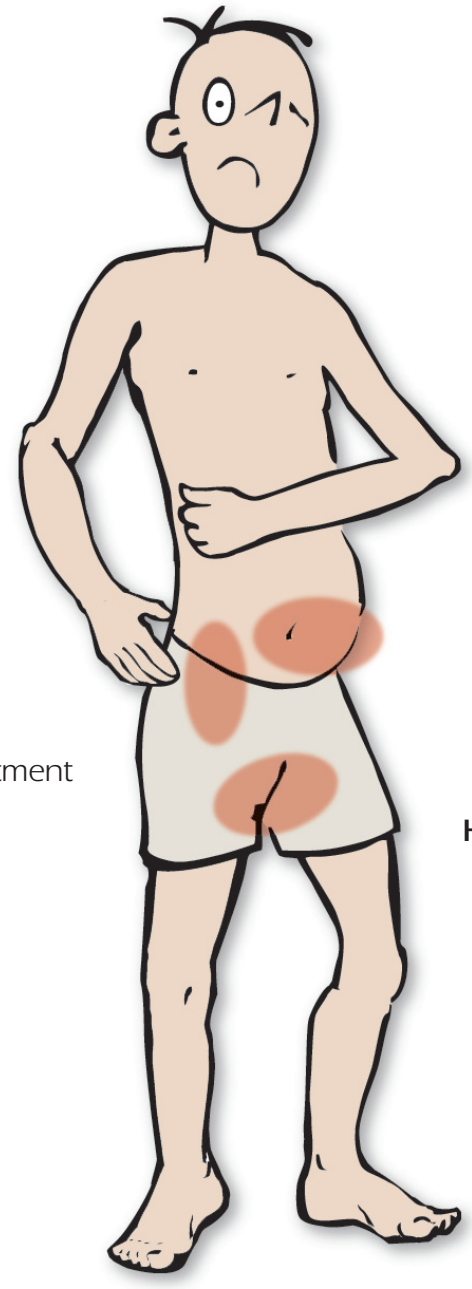
2. Avoid using a straw, chewing gum, and do not smoke. Negative pressure from the sucking action can dislodge the clot and aggravate the bleeding site.

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

4. If Desmopressin (DDAVP®, Ostostim®, or Stimate®) has been utilized for treatment, the patient has fluid restrictions for 24 hours.

Gastrointestinal / Urinary Tract Bleeding

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.

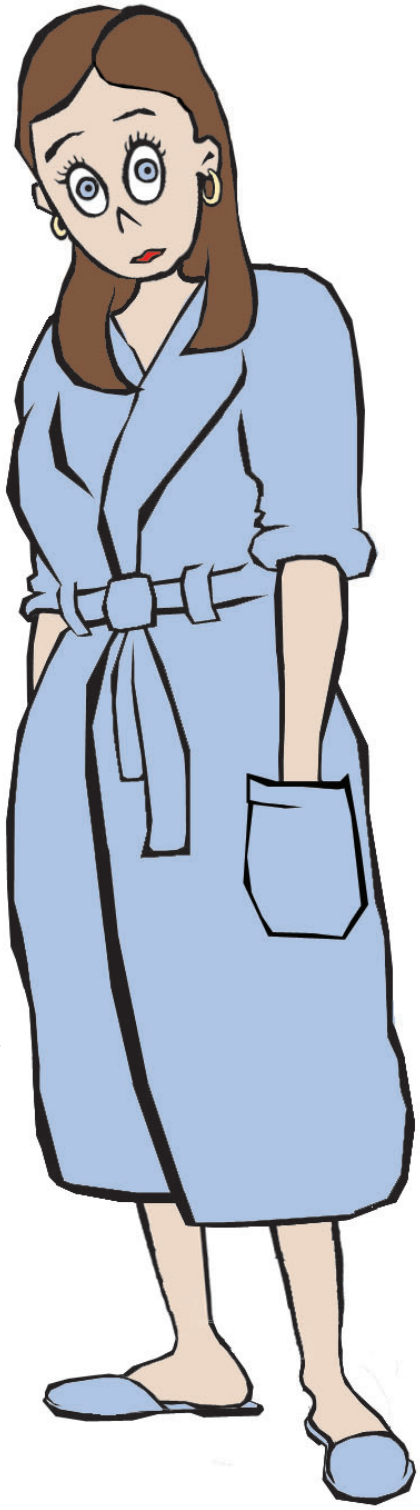
Genitourinary bleeding

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® 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®*) 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."
Kouides, P.A., Phatak, P. D., Burkart, P., et al. (2000). Gynecological and obstetrical morbidity in women with type 1 von Willebrand disease : results of a patient survey. Hemophilia, 6(6), 643-648.

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®*) Adjunctive treatment with intravenous or oral anti-fibrinolytics may be useful (see pg. 9).

Soft Tissue / Muscle / Joint Bleeding

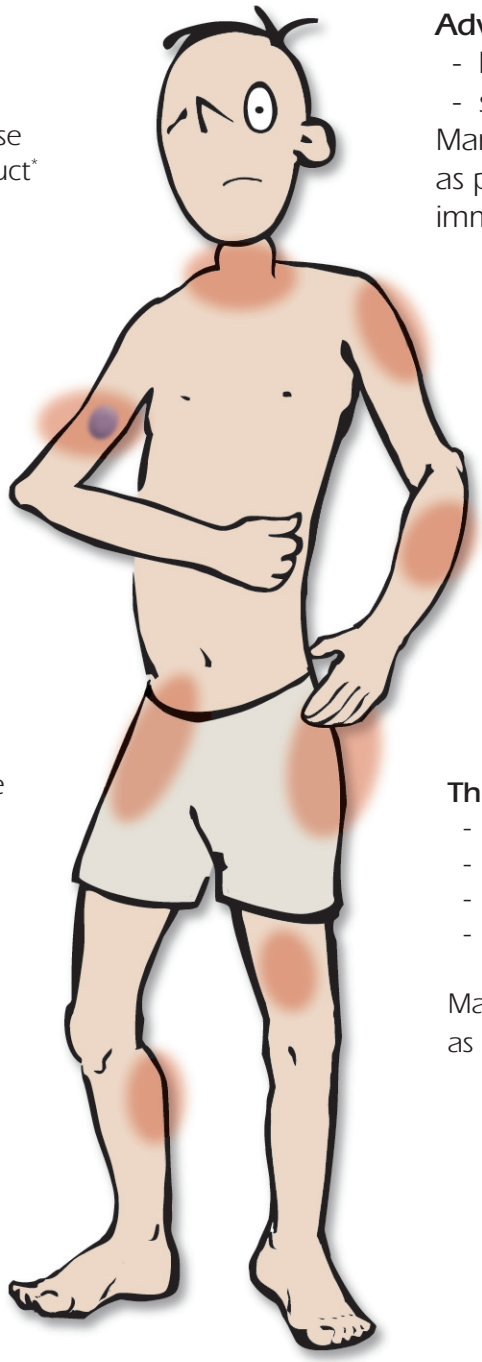
Early onset joint bleed
- tingling - pain
- limited range of motion

Advanced joint bleed
- heat - pain
- swelling

Management: Treatment product* as per hematologist. Ice and immobilization for comfort.

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.**

Deltoid / forearm bleed
- increased swelling and bruising
- observe for symptoms of compartment syndrome

Management: Treatment product* as per hematologist R.I.C.E.**

Iliopsoas bleeds
- flexed hip
- pain /inability to extend the leg on the affected side

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

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

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

DESMOPRESSIN - brand names: DDAVP®, OCTOSTIM®, STIMATE®

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®*.

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.

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	
Amicar 10% topical solution	Mix 2 ml Amicar (IV preparation 250 mg/ml), and 3 ml sterile water for injection Soak gauze in solution, squeeze out excess and apply to area. Discard solution after 24 hours
Tranexamic Acid 5% topical solution	Mix 5 ml Tranexamic acid, use the IV preparation 100mg/ml (5 ml ampule size) and 5 ml sterile water for injection Soak gauze in solution, squeeze out excess and apply to area. Discard solution after 24 hours

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.

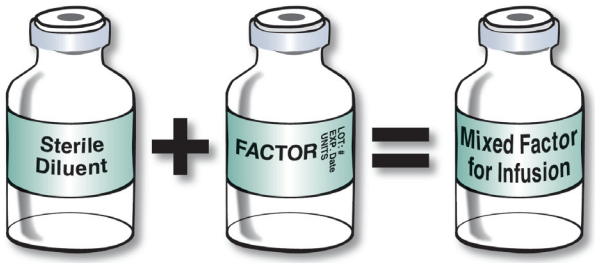
Topical agents such as topical Thrombin® and Gelfoam® and fibrin glue (Tisseal-R) may also be used to help control mucous membrane bleeding.

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®

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

Example for dose calculation

Patient's weight = 50 kilograms

Order: 60 ristocetin co-factor units/kg IV

60 ristocetin co-factor units X 50 kg =

3,000 ristocetin co-factor units

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.).

2625 I.U. **VWF :RCo/vial**
866 I.U. FVIII/vial
Expires: June 14, 2007
Lot Number: 22566611A

Information on front of Humate-P® box

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.

Pressure for a minimum of 5 minutes after a needle stick

Ice for 15 -20 minutes

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.

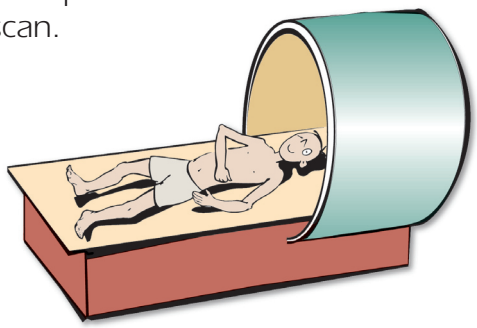
Invasive Procedures, Labs, X-rays

Treatment should never be delayed for laboratory studies to be drawn or completed.

Head injury

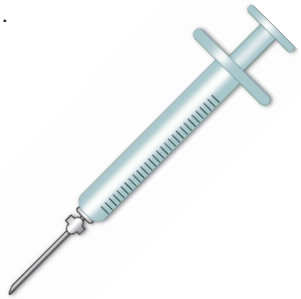
First give the recommended treatment*. . .

. . .then perform a CT scan.

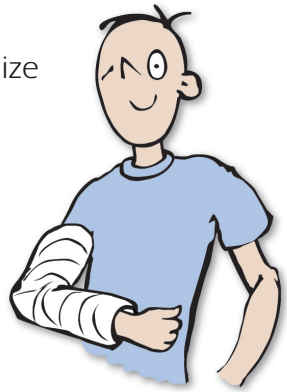


Fracture

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).

 *Recommended treatment table inside front cover

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

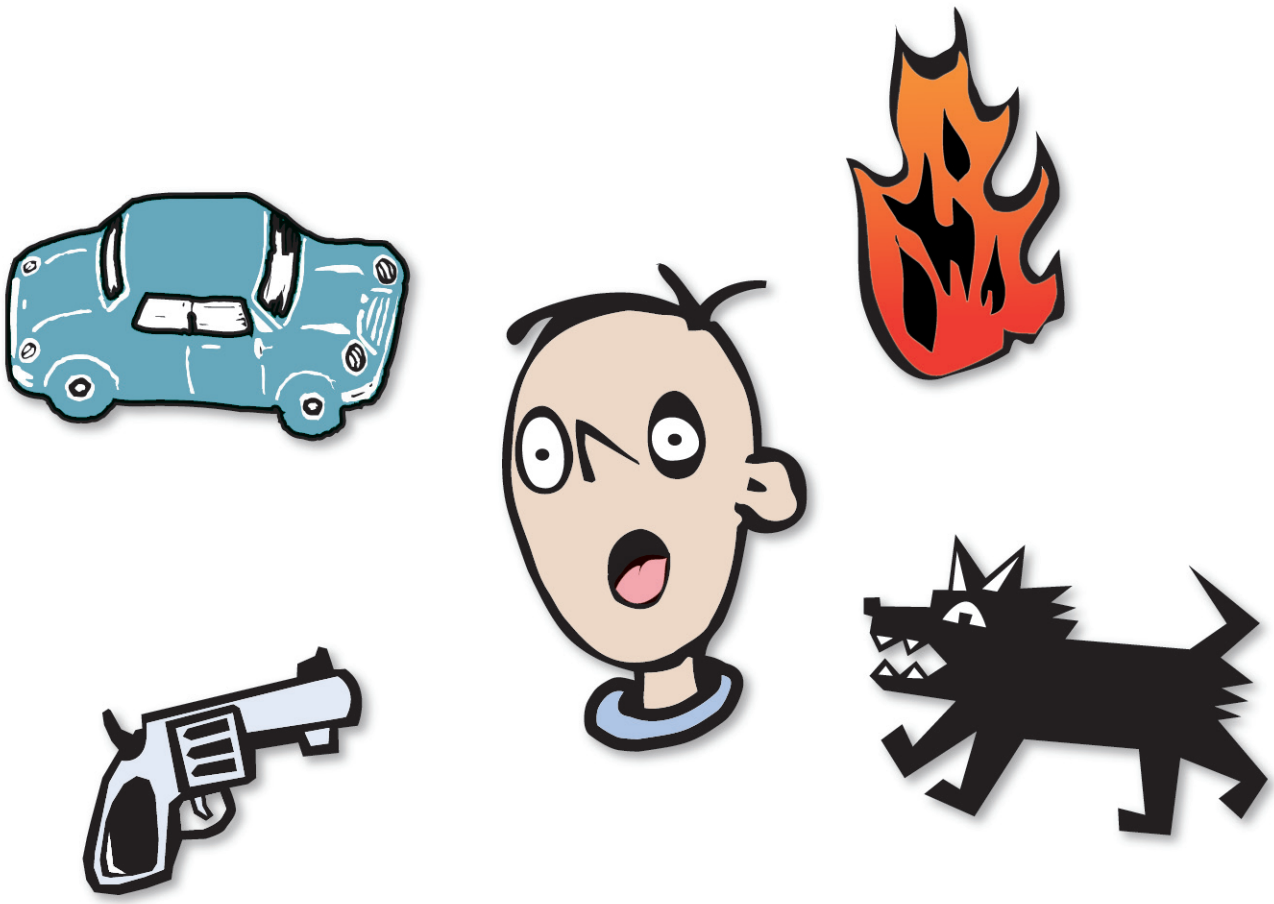
Invasive procedures should be performed as clinically indicated, i.e. lumbar puncture with symptoms of meningitis. However, factor replacement treatment* should be given prior to the procedure.

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.

Trauma / Emergencies



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®*) 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-fibrinolytics (pg. 9).

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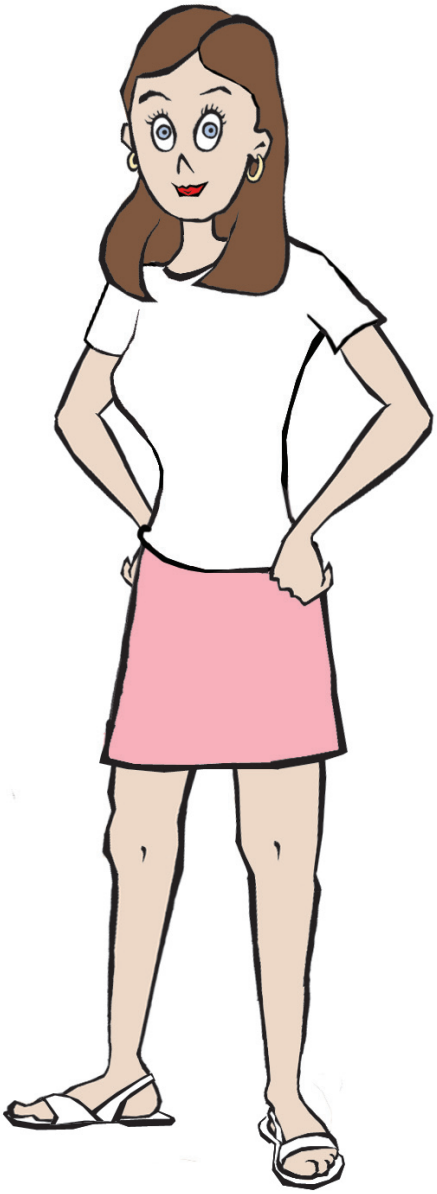

Sponsor Information

Emergency Care for Patients with von Willebrand Disease was made possible by a generous grant from the ZLB Behring Foundation for Research and Advancement of Patient Health.

ZLB Behring Foundation

Production Team - Project Manager: Art Gardner
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