Bleeding Disorders

Hemophilia

Genetic bleeding disorder more common in men
80% of Hemophilia are A (Factor VIII deficiency)
20% of Hemophilia are B (Factor IX deficiency)

Mild (6-25% of normal factor levels)
-rarely joint involvement
-may never have a bleed
-can bleed severely with injury, surgery, invasive procedures

Moderate (1-5% of normal factor levels)
-some joint involvement
-bleed about 1 x month
-can bleed with even the slightest injury

Severe (less than 1% of normal factor levels)
-if untreated, will have joint involvement
-if untreated, will bleed a few times a week
-if untreated, can bleed without injury

Treatment
Minor cuts and scrapes: compression and bandages
More traumatic injuries: factor replacement therapy (from plasma or synthetic recombinant factor) is based on a patient's weight and can be thousands of dollars per infusion. Encourage regular exercise & being fit. Patient may have disabilities which affect appointment reliability and oral hygiene.

Inhibitors (more common in African American decent)
30% of severe Hemophilia A patients and 4% of severe Hemophilia B patients, it's usually detected in childhood, but can happen anytime. **Consultation with hematologist is critical before dental treatment**.
Treatment of choice:
-multiple doses of Recombinant Factor 7a
Low Responding Inhibitor Therapy choices:
-greater amount of factor and additional doses of factor
-"Immune Tolerance" daily factor infusions to overcome body's immune system antibody production
High Responding Inhibitor Therapy choices:
-plasmapheresis in a hospital
-prothrombin concentrates (lack of efficacy, thrombosis risk, stimulation of more inhibitor possible) ***avoid using antifibrinolytics on prothrombin patients**

von Willebrand Disease

Genetic bleeding disorder in men and women. Deficient von Willebrand proteins disrupt platelet aggregation and make Factor 8 unstable, leading to superficial bleeding like bruising, nose bleeds, bleeding gums, heavy menstrual bleeds, bleeding after surgery.

Type I deficient *quantity* of vW proteins, most common form, not many bleeds other than from surgery.

Type II deficient *quality* of vW proteins (IIA abnormal molecular weight disrupts aggregation, IIB spontaneous aggregation/thrombocytopenia, IIM deficient platelet receptors disrupt aggregation, IIN rapid proteolysis of Factor 8 similar to Hem A).

Type III total deficiency of vW proteins, most serious and most rare, can have joint involvement.

Treatment
DDAVP/desmopressin acetate: a synthetic hormone that stimulates release of vW proteins with Factor 8, so it won't work in Type III patients and is **contraindicated in Type IIB patients** (consult with hematologist first). It's available via injection or nasal spray.
Factor replacement therapy: cryoprecipitate or fresh frozen plasma, but factor may be destroyed.

Platelet Dense Granule Deficiency

Deficiency in functioning platelet dense granules affects the release of ADP for platelet aggregation and Calcium for clotting cascade, causing easy bruising, nose bleeds, bleeding from the gingiva, heavy or prolonged menstrual bleeding, bleeding after childbirth, and abnormal bleeding after surgery, circumcision, or dental work.

Treatment
Antifibrinolytic drugs, Desmopressin, and Platelet Transfusions are used for surgical procedures, dental work, injuries, and trauma.

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Quick Reference Guide

Bleeding Disorders

Risk Prevention:

Establish Dental Home
Injury Prevention
Anticipatory Guidance
Oral Hygiene
Patient Education

Be Prepared:

Fill out referral form
Know Hematologist's recommendations
Know if patient followed recommendations

"Safe": examination, gentle radiography, fluoride application, sealants, probing, restorations, impressions, dentures.

Pretreatment Needed: oral surgery, periodontal surgery, extractions, and implants.

Take These Precautions:

Use gingival retractions cords
Avoid wooden wedges and matrix bands
Use rubber dams
Careful placing rubber dam clamp
Careful with tori for radiographs
Careful with impression trays
Careful use of saliva ejectors
Wet cotton rolls for removal
Watch out for frenum
Endo preferred over extractions
Sever PDL for extractions
Group extractions together
Figure 8 suturing
Dissolvable sutures
Gel foam sponges with thrombin powder/sterile water
Post op stent with a well for hemostatic agent delivery
(for a stent= no brushing or flossing for 2-3 days and only drink liquids for 48 hours)
Fibrin glue (synthetic thrombin and fibrinogen)
30 gauge short needle for local anesthetic
Aspirate for all injections (**IF POSITIVE TELL PATIENT**)
Avoid mandibular blocks and lingual infiltrations
Try buccal infiltration with Articaine
Try intraligamental or interosseous techniques
Palatal and intrapapillary techniques are safe
Avoid children being numb for too long of time
Low dose acetaminophen is ok
Avoid all aspirins and COX-1 NSAIDS
Celebrex is a good alternative to ibuprofen

To Control a Bleed:

Consult with Hematology Treatment Center
Apply pressure
Try topical agents
Administer treatment after a consultation

Topical Agents

Gelfoam with thrombin powder and sterile water
Fibrin sealant (glue)
Bovine thrombin
Collagen sponge
30% aluminum chloride gel with irrigation after use

Treatments

DDAVP/desmopressin acetate (Stimate™)
Useful for mild Hem A, may work for Type I von Willebrand, won't work for Type III vW or Hem B, contraindicated for Type IIB vW. Side effects include transitory facial flushing, increased heart rate, and change in blood pressure. **Avoid** taking in more than 1.5 liters of fluid for 24hrs after taking it. **Avoid** use in patients with electrolyte imbalances, cystic fibrosis, heart failure, renal disorders, birth control pills, and elders at risk for cerebrovascular events or cardiovascular events.

- 0.3mcg/kg given intravenously in 30-50ml normal saline over 30 minutes
- daily dose of 150mcg per nasal puff (0.1ml of 1.5mg/ml solution) give 1 puff for person < 50kg and 2 puffs (one per nostril) for person >50kg **Avoid** giving 10mcg per puff formulation
- If still bleeding after 3 days, factor concentrate may be necessary so consult your HTC

Antifibrinolytics
Tranexamic acid

- buy 30 tab bottle of 650mg tabs, crush 2 tabs into normal saline (5% solution) swish and swallow/spit q 6 hrs, keep in dark refrigerator for up to 5 days
- intravenously 10mg/kg q 8hrs
- take 2 650mg tabs, 3 x day for 5-7 days

Aminocaproic acid (Amicar™)

- 50-60mg/kg adults and children, give 4-5g orally 1 hr before dental procedure, then 1g/hr or 4-6g q 4-6hrs for 5-7 days post-op
- **Avoid** giving more than 24g in 24hrs
- If still bleeding after 8-12 hours, an infusion of factor may be necessary so consult your HTC

Factor Concentrates

- Available to the patient through their HTC