NIÑO QUE SANGRA

- **DR: GUSTAVO CEA SANHUEZA.**
- **PEDIATRÍA HBV-UACH**

SANGRADO ANORMAL

- *Duración o volumen desproporcionado al evento*.

- Epistaxis > de 15 minutos.
- Persiste al día siguiente del dentista.
- Menstruaciones de mas de 7 días.
- Equimosis de tamaño mayor a una naranja.
- Hemorragia en lactantes con vacunas o punciones.
- Equimosis o hematomas desproporcionados al iniciar el gateo .
- Equimosis en lugares distintos de la frente, codos y pretibiáles.

DOGMA 1

- *Cuando un niño ha sido sometido a alguna cirugía o extracciónes dentales sin complicaciones hemorrágicas es muy improbable que tenga un trastorno hemorrágico congénito primario.*

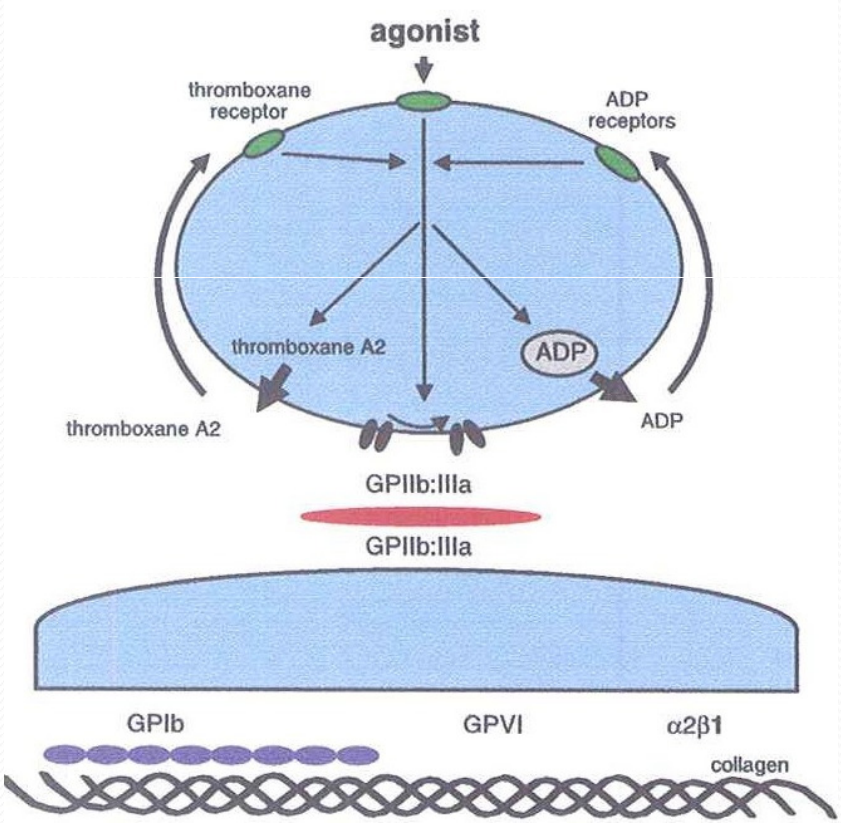
DOGMA 2

- *Los hematomas musculares profundos y las hemartrosis en los niños son siempre anormales. Con o sin traumatismo.*

DOGMA 3

- *El hallazgo de equimosis antes de la edad de gateo o en sitios inhabituales debe alertar por posible maltrato.*

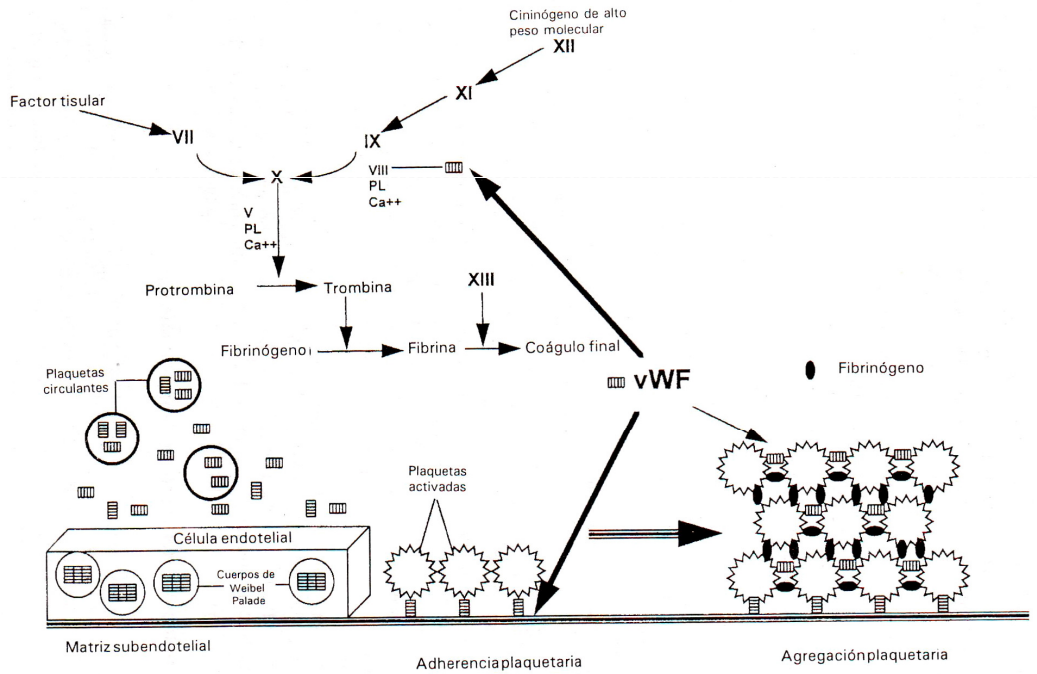
ALTERACIONES DE LA HEMOSTASIA PRIMARIA



TIPO DE SANGRADO.

- Púrpura cutáneo mucoso.
- Sangrado mucoso.
  - Epistaxis.
  - Hemorragia gingival.
  - Menorragia.
- Sangrado persistente.

ENFERMEDAD DE VON WILLEBRAND



FWD CLASIFICACIÓN.

Table 3 Classification of von Willebrand disease based on specific von Willebrand factor tests

VWD type	VWF:Ag	VWF:RCo	RCo/Ag	FVIII	Multimer pattern	Low-dose RIPA	Platelets
1	↓	↓	=	N or ↓	N	Absent	N
2A	↓	↓	RCo<Ag	N or ↓	Abnormal	Absent	N
2B	↓	↓	RCo<Ag	N or ↓	Abnormal	↑	N or ↓
2M	↓	↓	RCo<Ag	N or ↓	N	Absent	N
2N	N or ↓	N or ↓	=	<30 IU/dL	N	Absent	N
3	Absent	Absent	NA	<10 IU/dL	Absent	Absent	N

RIPA ristocetin-induced platelet aggregation, *VWF* von Willebrand factor, *VWD* von Willebrand disease, *NA* not applicable, *N* normal, *VWF:Ag* VWF antigen assay, *VWF:RCo* VWF ristocetin cofactor activity, *FVIII* factor VIII

TROMBOCITOPENIA

Table 1 Classification of thrombocytopenia in the neonate and child

Neonatal thrombocytopenia	Childhood thrombocytopenia
Early onset (<72 h)	Destructive thrombocytopenia
Placental insufficiency	Immune thrombocytopenia
Perinatal asphyxia	Medication
Perinatal infection	Infection
Disseminated intravascular coagulation	Hemolytic uremic syndrome
Alloimmune	Thrombotic thrombocytopenic purpura
Autoimmune	Catheter-related thrombosis
Congenital infection	Heart disease
Thrombosis	Disseminated intravascular coagulation
Bone marrow infiltration replacement	Von Willebrand disease type 2B
Kasabach–Merritt syndrome	Kasabach–Merritt syndrome
Metabolic disease	Hypersplenism
Congenital/inherited	Hypothermia
	Massive bleeding
Late onset (>72 h)	
Sepsis	Decreased production
Necrotizing enterocolitis	Inherited (large/normal/small platelets)
Congenital infection	Congenital aplastic anemia
Autoimmune	Congenital amegakaryopoeiseis
Catheter-related thrombosis	Bone marrow infiltration
Kasabach–Merritt syndrome	Vitamin B_{12} deficiency
Metabolic disease	Folic acid deficiency
Medication	Acidosis
Congenital	Infection
Inherited (large/normal/small platelets)	Medication

TROMBICITOPENIA NN PRECOZ (<72 h)

- Insuficiencia placentaria.
- Asfixia perinatal.
- Infecciones perinatales.
- CID.
- Aloinmune.
- Autoinmune.
- Congénita hereditaria.

TROMBOCITOPENIA NN ALOINMUNE.

- 1: 1000 a 1500 embarazos.
- IgG anti HPA-1a en 75%.
- Plaquetas < 20.000.
- HIC en 10 a 20% sin tratamiento
- RNT en lo demás sano.
- Plaquetas normales en la madre.

TROMBOCITOPENIA NN TARDIA (>72 h)

- Sepsis.
- Enterocolitis necrotizante.
- Infecciones congénitas.
- Autoinmune.
- Asociada a cateter.
- Enf. Metabólicas.
- Kasabach-Merrit.

TROMBOCITOPENIA DEL NIÑO

- *Por disminución de la producción:*
  - Hereditarias.
  - Anemia aplástica.
  - Infiltración medular.
  - Déficit de B12 o foláto.
  - Infecciones.
  - Medicamentos.

TROMBOCITOPENIAS DEL NIÑO.

- *Por destrucción excesiva*:
  - PTI
  - Infecciones.
  - SHU.
  - CID.
  - Hiperesplenismo.

PTI

- Plaquetas < 100.000.
- Manifestación significativa en 3%.
- Riesgo con plaquetas < 10.000.
- HIC 0.1% a 0.3%.
- Crónico > 12 meses.
- Muy agudo y severo pero autolimitado.

DISFUNCIONES PLAQUETARIAS.

- *Adquiridas.*
  - Medicamentos.
  - Insuficiencia renal.
  - Insuficiencia hepática.
  - Enfermedades neoplásicas.

DISFUNCIONES HEREDITARIAS.

- 1 a 2 casos por millón de niños.
- Solo en 50% → diagnóstico preciso.
  - Glanzmann 32%
  - Aspirin-like 21%
  - Receptores 17%
  - Storage pool 15%
  - B. Soulier 8%

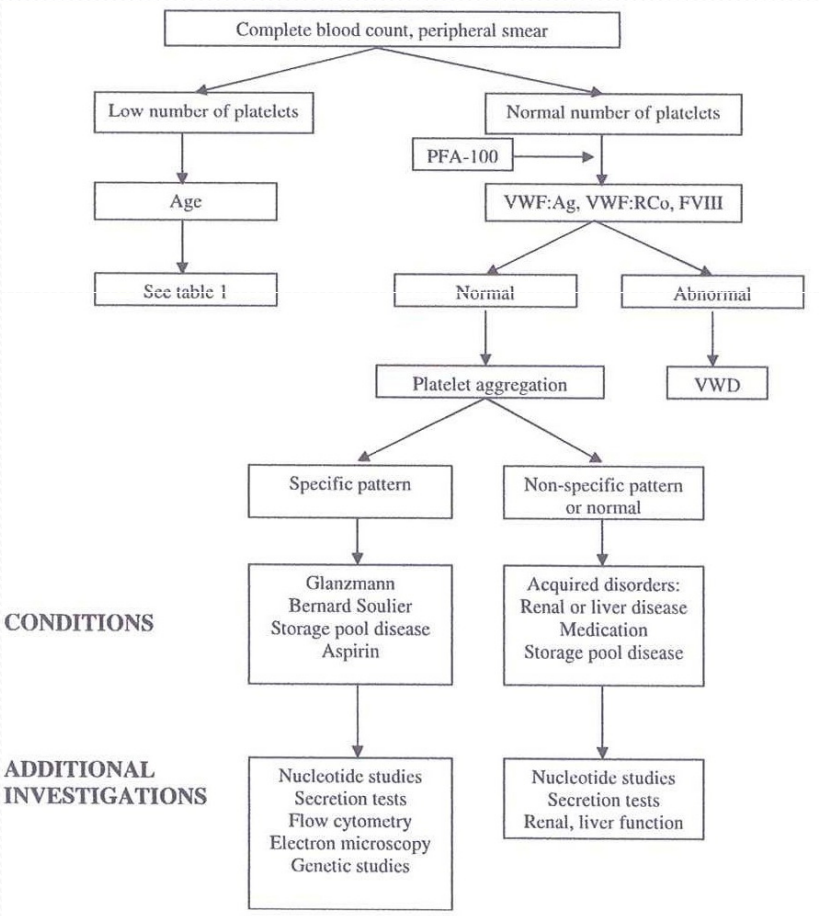
DISFUNCIONES CLASIFICACION.

Table 4 Characteristic findings on light transmission aggregometry

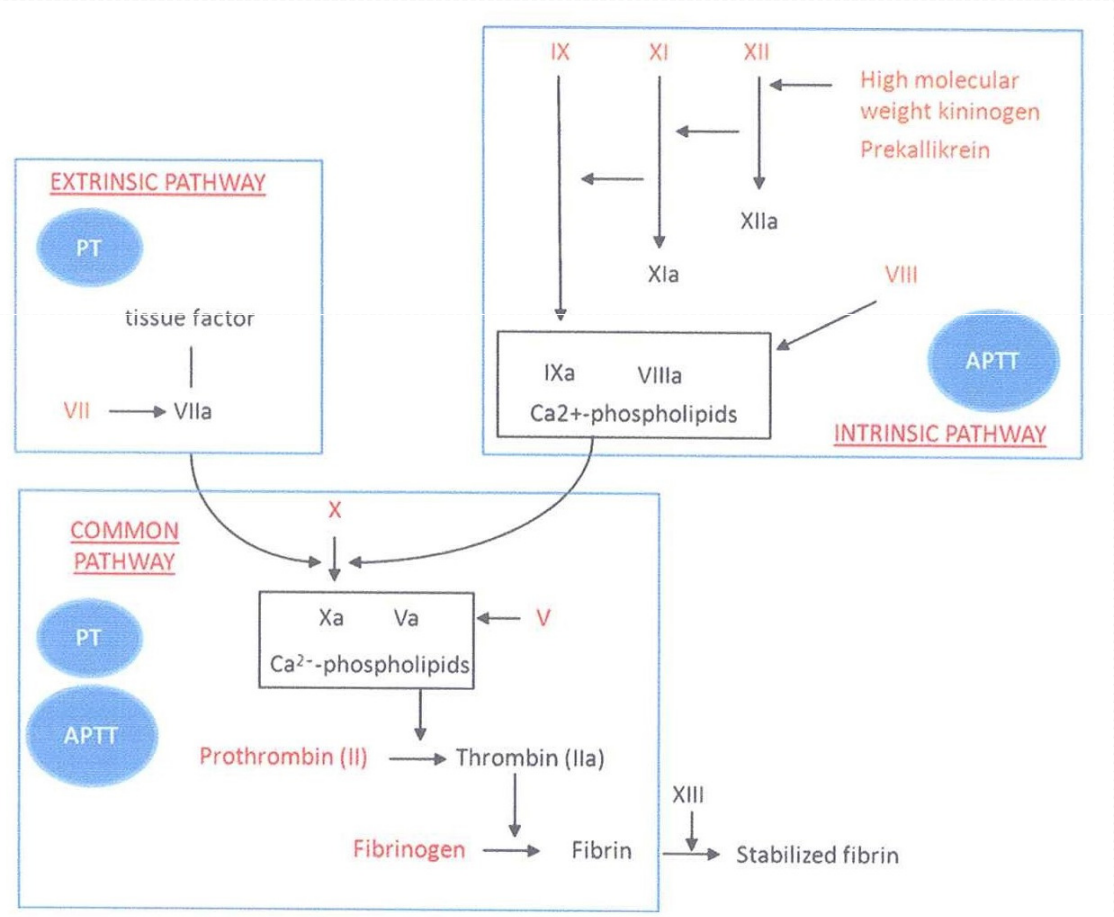
		Platelet aggregation				
Disorder	Platelet morphology	Epinephrine	ADP	Collagen	Ristocetin	AA
Glanzmann	Normal	−	−	−	+	−
Bernard Soulier	Giant platelets	+	+	+	−	+
Aspirin	Normal	+	+	±	+	−
Storage pool disease						
Gray platelet syndrome	Large with decreased α-granules	+	−	−	+	+
δ-Granules deficiency	Decreased δ-granules	−	±	−	±	±

AA arachidonic acid, *ADP* adenosine 5′-diphosphate

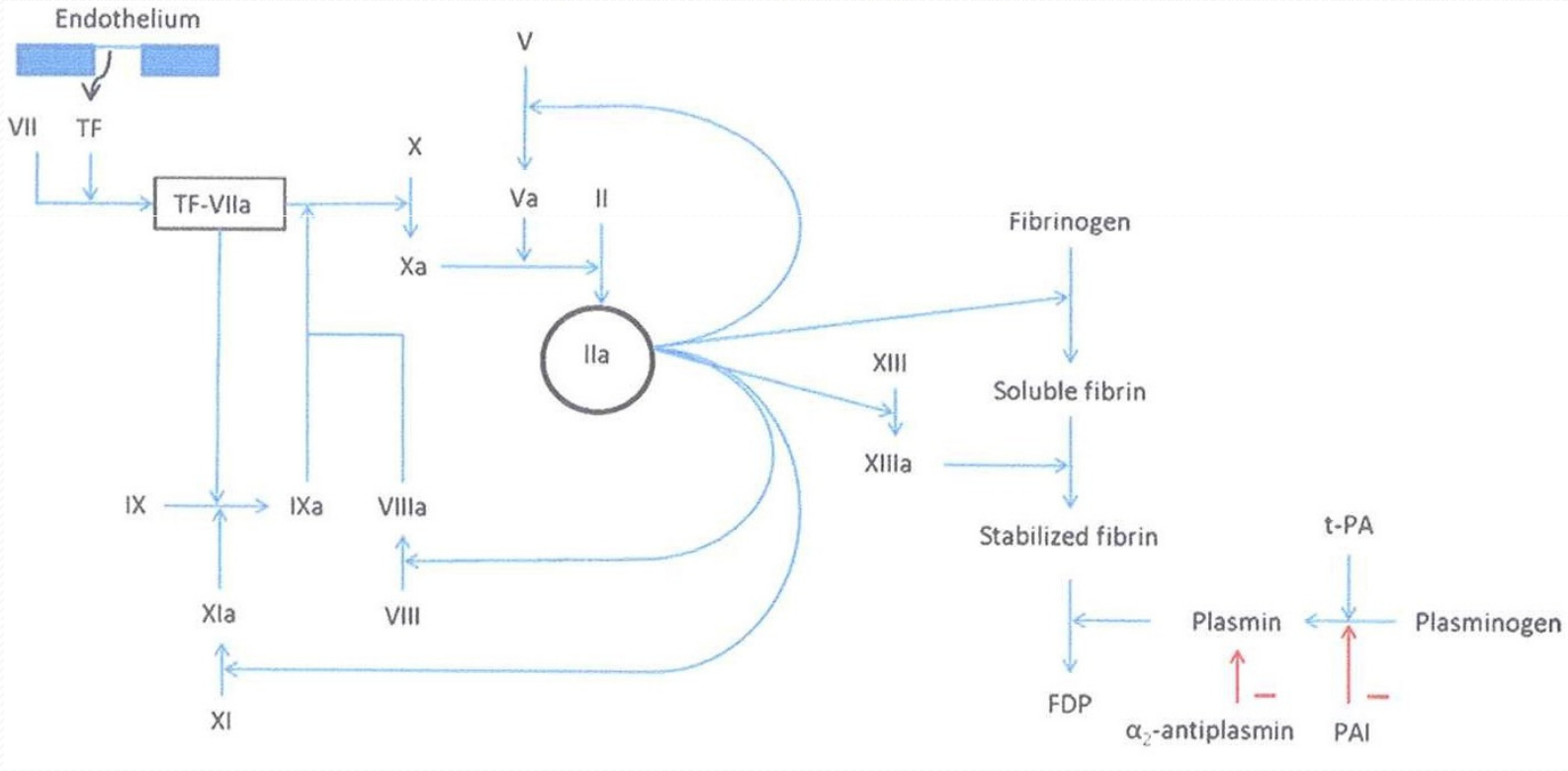
ALGORITMO


Complete blood count, peripheral smear
Low number of platelets
Normal number of platelets
PFA-100
Age
VWF:Ag, VWF:RCo, FVIII
See table 1
Normal
Abnormal
Platelet aggregation
VWD
Specific pattern
Non-specific pattern or normal
CONDITIONS
Glanzmann
Bernard Soulier
Storage pool disease
Aspirin
Acquired disorders:
Renal or liver disease
Medication
Storage pool disease
ADDITIONAL INVESTIGATIONS
Nucleotide studies
Secretion tests
Flow cytometry
Electron microscopy
Genetic studies
Nucleotide studies
Secretion tests
Renal, liver function

ALTERACIONES DE LA HEMOSTASIA SECUNDARIA.



MODELO ACTUAL.



TIPO DE SANGRADO.

- *Hemorragias profundas:*

  - Hemartrosis.
  - Hematomas musculares.
  - HIC.

- Sangrado tardío.

HEMOFILIAS A Y B

- 1:5000 y 1:30000 hombres.
- 30% mutaciones "*de novo*"
- Severidad según nivel de factor:
  - Severa < 1%
  - Moderada 1 a 5 %
  - Leve 6 a 40%

OTROS TRASTORNOS HEREDITARIOS.

Table 1 Incidence, inheritance, clinical manifestations, and therapeutic modalities, for both the congenital coagulation factor deficiencies and the fibrinolytic defects [4, 6, 15]

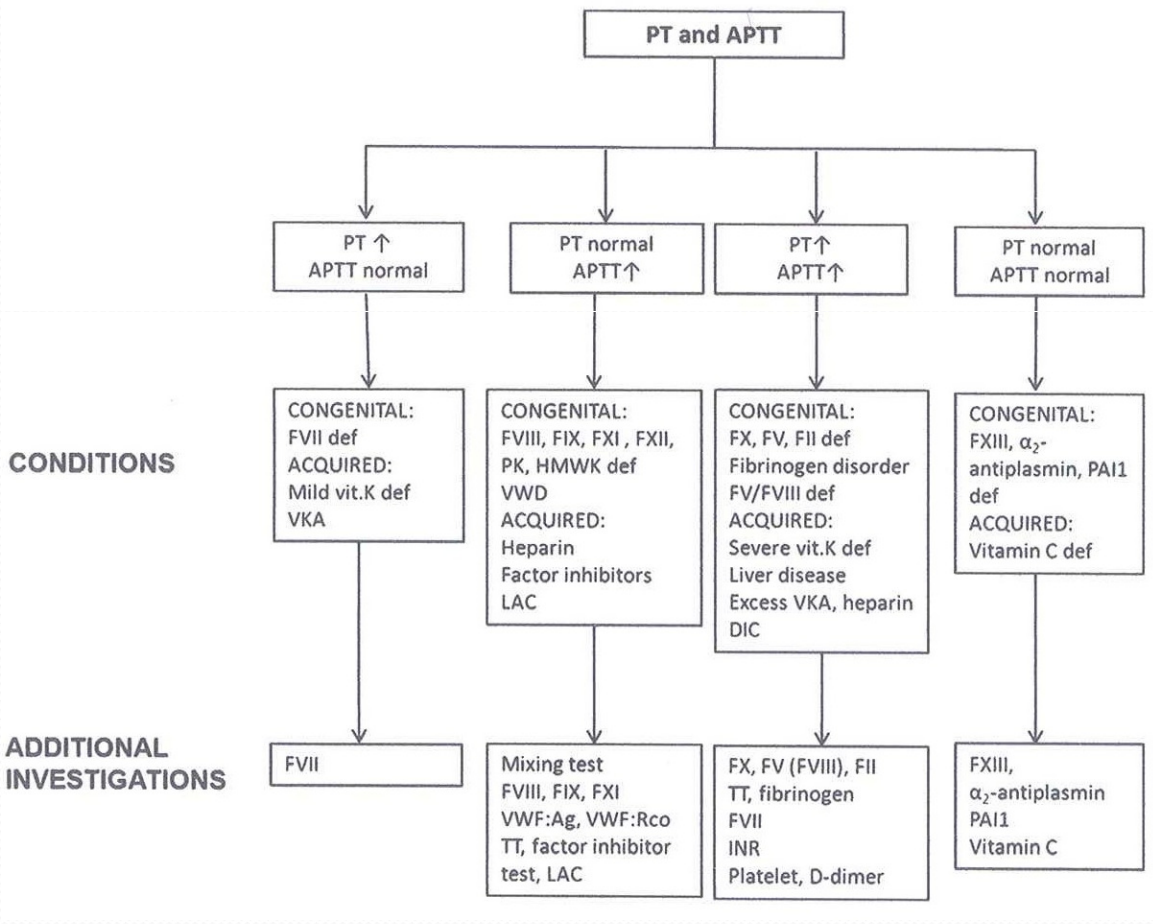
	Incidence	Inheritance	Clinical manifestations	Therapeutic modalities
Congenital coagulation factor deficiencies				
Hypo-, or afibrinogenemia (FI deficiency)	1 per 1,000,000	AR	Bleeding of umbilical cord, in gastro-intestinal tract, in genito-urinary tract, in CNS, posttraumatic/post-surgery bleeding, mucocutaneous bleeding or joint bleeding (rare)	Pd-fibrinogen conc.Plasma
FII deficiency (prothrombin deficiency)	1 per 2,000,000	AR	Bleeding of muscle or joint, mucocutaneous bleeding, or CNS bleeding (rare)	Prothrombin complex conc. Plasma
FV deficiency	1 per 1,000,000	AR	Mucocutaneous bleeding, joint bleeding (rare) or umbilical cord bleeding (rare)	Plasma
FVII deficiency	1 per 500,000	AR	Bleeding of joint, CNS bleeding or mucocutaneous bleeding	Pd-FVII conc.Rec. FVIIa conc.
FVIII deficiency (hemophilia A)	1 per 5,000 males	X-linked	Bleeding of joint or muscle, in CNS, posttraumatic/ post-surgery bleeding or mucocutaneous bleeding	Rec. FVIII conc.Pd-FVIII conc.
FIX deficiency (hemophilia B)	1 per 30,000 males	X-linked	Bleeding of joint or muscle, in CNS, posttraumatic/ post-surgery bleeding or mucocutaneous bleeding	Rec. FIX conc.Pd-FIX conc.
FX deficiency	1 per 1,000,000	AR	Mucocutaneous bleeding, posttraumatic/post-surgery bleeding, umbilical cord or joint bleeding (rare)	Prothrombin complex conc. Plasma
FXI deficiency (hemophilia C)	1 per 1,000,000	AR	Posttraumatic bleeding/post-surgery bleeding, mucocutaneous bleeding	Pd-FXI conc.Plasma
FXII deficiency	25 per 1,000	AR	None	Not necessary
FXIII deficiency	1 per 1,000,000	AR	Bleeding of umbilical cord or in CNS, poor wound healing	Pd-FXIII conc. or plasma
Congenital fibrinolytic defects				
Antiplasmin deficiency	Rare	AR	Mucocutaneous bleeding, posttraumatic/post-surgery re-bleeding, bleeding of joint, umbilical cord or in CNS	Antifibrinolytic drugsPlasma
Plasminogen activator inhibitor 1 deficiency	Rare	AR	Mucocutaneous bleeding, posttraumatic/post-surgery re-bleeding, bleeding of joint or in CNS	Antifibrinolytic drugs

FV factor V, *AR* autosomal recessive, *CNS* central nervous system, *conc.* concentrate, *Pd* plasma-derived, *rec.* recombinant

TRASTORNOS ADQUIRIDOS.

- Déficit de vitamina K.
- Anticoagulantes.
- CID.
- Enfermedad hepática severa.

ALGORITMO.


PT and APTT
PT ↑
APTT normal
PT normal
APTT↑
PT↑
APTT↑
PT normal
APTT normal
CONDITIONS
CONGENITAL:
FVII def
ACQUIRED:
Mild vit.K def
VKA
CONGENITAL:
FVIII, FIX, FXI , FXII,
PK, HMWK def
VWD
ACQUIRED:
Heparin
Factor inhibitors
LAC
CONGENITAL:
FX, FV, FII def
Fibrinogen disorder
FV/FVIII def
ACQUIRED:
Severe vit.K def
Liver disease
Excess VKA, heparin
DIC
CONGENITAL:
FXIII, α2-antiplasmin, PAI1 def
ACQUIRED:
Vitamin C def
ADDITIONAL INVESTIGATIONS
FVII
Mixing test
FVIII, FIX, FXI
VWF:Ag, VWF:Rco
TT, factor inhibitor test, LAC
FX, FV (FVIII), FII
TT, fibrinogen
FVII
INR
Platelet, D-dimer
FXIII,
α2-antiplasmin
PAI1
Vitamin C

FIN