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AN APPROACH TO

BLEEDING DISORDERS
NORMAL HEMOSTASIS

After injury, 3 processes halt bleeding

- Vasoconstriction
- Platelet Plug formation
- Activation of coagulation cascade
Disorders of Hemostasis fall into 3 groups

- Platelet disorders
- Disorders of coagulation cascade
- Vascular disorders
A) Platelet Disorders

1) Decreased platelet survival

- Idiopathic thrombocytopenic Purpura
- Viral infections
- DIC
- Drugs i.e. Aspirin, NSAIDS
- TTP
- Hypersplenism
- Autoimmune thrombocytopenia
2) Decrease platelet production

- Aplastic anemia
- Lymphoma/Leukemia
- Myelofibrosis
- Myelodysplasia
- Megaloblastic anemia
3) Decreased platelet function

- Platelet function disorders
- Chronic renal failure
B) Disorders of Coagulation Cascade

1) Congenital
   - Hemophilia
   - Von Willibrand Disease
   - Other factors Deficiency

2) Acquired
   - Hemorrhagic Disease of Newborn
   - Anti coagulants; Heparin, Warfarin
   - Liver disease
   - DIC
   - Malabsorption Syndrome
C) Vascular Disorders

- Henoch schonlein Purpura
- Ehlers Danlos syndrome
- Steroids
- Trauma / Pressure
- Scurvy
- Severe malnutrition
- SLE
Idiopathic Thrombocytopenic Purpura

Commonest cause of acute onset of thrombocytopenia
1-4 weeks after exposure to common viral infections

Clinical Feature
Petechiae / Purpura
Bleeding from gums & mucous membranes
Intracranial bleed < 1%
Splenomegaly rare
Chronic ITP associated with SLE
Purpura rash on the forearm
Diagnosis

Platelet count < $20 \times 10^9/L$
Megakaryocytosis on bone marrow biopsy

Treatment

Prednisolone
Intravenous immunoglobulin (IVIG)
IV Anti D therapy
Splenectomy
  Older children > 4 yr
Chronic ITP
Intracranial hemorrhage
Hemolytic Uremic Syndrome

Triad of

Microangiopathic hemolytic anemia
Thrombocytopenia
Acute Renal failure

Follows acute gastroenteritis caused by E-coli 0157:H7

Treatment

Supportive care
Fluid management
Plasmapheresis, if CNS complications
Thrombotic Thrombocytopenic Purpura

Acquired deficiency of metalloproteinase

Pentad of;

Fever
Microangiopathic hemolytic anemia
Thrombocytopenia
Abnormal renal function
CNS change
Treatment

Plasmapheresis
Corticosteroids
Splenectomy
Congenital Thrombocytopenia Syndromes

1) Congenital Amegakaryocytic Thrombocytopenia

Presents by 3-4 wks of life
Petechiae / Purpura
Absence of megakaryocytes on bone marrow
Bone Marrow Transplantation
2) TAR Syndrome (Thrombocytopenia Absent Radius)

*Skeletal Abnormality

*Spontaneous remission of thrombocytopenia in first few years of life
Thrombocytopenia Absent Radius Syndrome
3) Wiskott Aldrich Syndrome

* X linked recessive
* Thrombocytopenia, eczema, recurrent infection
* Bone marrow shows normal no of megakaryocytes with abnormal bizarre morphology
* Treatment ; Splenectomy, BM transplant
1) Bernard-Soulier Syndrome

* Autosomal recessive
* Thrombocytopenia, giant platelets, prolonged bleeding time > 20sec
* Absence of VWF receptor on platelet membrane
* Absent ristocetin-induced platelet aggregation
2) Glanzmann Thrombasthenia

* Autosomal recessive
* Deficiency of platelet fibrinogen receptor on platelet surface
* Abnormal aggregation with all agonist except ristocetin
Treatment of Platelet function disorders

* Desmopressin
* Platelet transfusion
* Recombinant Factor VIIa
* Stem cell transplantation
Vascular Disorders

1) Henoch Schonlein Purpura

- Palpable Purpura on lower extremities & buttocks
- Arthritis
- Abdominal pain
- Renal involvement
- Purpura symmetric and on legs and buttocks
- Henoch Schonlein Purpura
Lab

Normal coagulation studies & platelet count
Pathological lesions ; leucocytoclastic angitis

Treatment

Supportive
Steroids
2) Ehlers Danlos Syndrome

* Disorder of collagen structure
* Easy bruising, poor wound healing
* Soft, hyperelastic skin & laxed joints that are easily subluxed
* Associated with sudden rupture of viscera
* Bleeding time mildly prolonged
* Platelets show defective aggregation to collagen
3) Acquired vascular disorders

*Scurvy
*Chronic corticosteroid therapy
*Severe malnutrition
*SLE
Clinical Evaluation --- Bleeding Disorder

**History**

Age of onset

*Congenital, Acquired*

Site of bleeding

*Mucus membrane – Platelets Disorders*

*Deep – Coagulation Disorders*

Bleeding stops and resumes

*Coagulation Disorders*
Profuse bleeding from superficial cut

*Platelets Disorders*

**History of fever**

*Infection*

**History of fever and neurological complications**

*Meningococcemia, TTP*

**Viral prodrome**

*ITP, HUS*

**Drug History**

**History of Trauma**
Repeated episodes of bleeding gums, prolong bleeding from cuts and massive bleeding from surgical procedures

*Congenital Disorders*

Bleeding from circumcision site or large hematomas at vaccination site

*Congenital Disorders*

Family history of easy bruising or abnormal bleeding

*Congenital Disorders*
Physical Examination

Large ecchymoses or hemarthroses

*Congenital Disorders*

Petechiae or Ecchymoses

*Platelets Disorders*

Purpura symmetric and on legs and buttocks

*HSP*

Extensive ecchymotic lesions in various stages

*Physical Abuse*
Palpable Purpura with history of minimal trauma

*Congenital Disorders, Platelets Disorders*

Umbilical stump bleeding

*Factor XIII deficiency*

Lymphadenopathy/Hepatosplenomegaly

*Leukemia, Lymphoma, Infections*

Jaundice and bruising

*Hepatic disease*
Lab Studies

- Low platelets, Normal PT and APTT
  *Thrombocytopenia*

- Normal platelet, PT and APTT, Abnormal Bleeding Time
  *Platelet function disorder*

- Normal platelet, PT and Bleeding Time but a prolong APTT
  *Hemophilia A & B*

- Normal platelet, PT, but prolong Bleeding Time & APTT
  *von Willebrand Disease*
- Normal platelet, APTT, and Bleeding Time, but a prolong PT
  \textit{Factor VII deficiency}

- Normal platelet, PT, APTT and Bleeding time
  \textit{Vascular disorder, Factor XIII deficiency}

- Normal platelet but a prolong PT and APTT
  \textit{CLD, Vitamin K deficiency}
- Normal PT and APTT, but a low platelet and Prolong Bleeding Time
  
  **vWD (type 2B), Bernard–Soulier Disease**

- Markedly low and small platelets
  
  **Wiskott-Aldrich syndrome**

- Markedly low platelet and large platelets
  
  **ITP, Bernard-Soulier Disease**

- Low platelet, prolong PT and APTT, Schistocytes
  
  **DIC, Microangiopathic Process**
Thank You