12.ULUSAL ACİLTIP KONGRESİ
SUENO DELUXE OTEL
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ATUDER
Acil Tıp Uzmanları Derneği

BLEEDING CHILD

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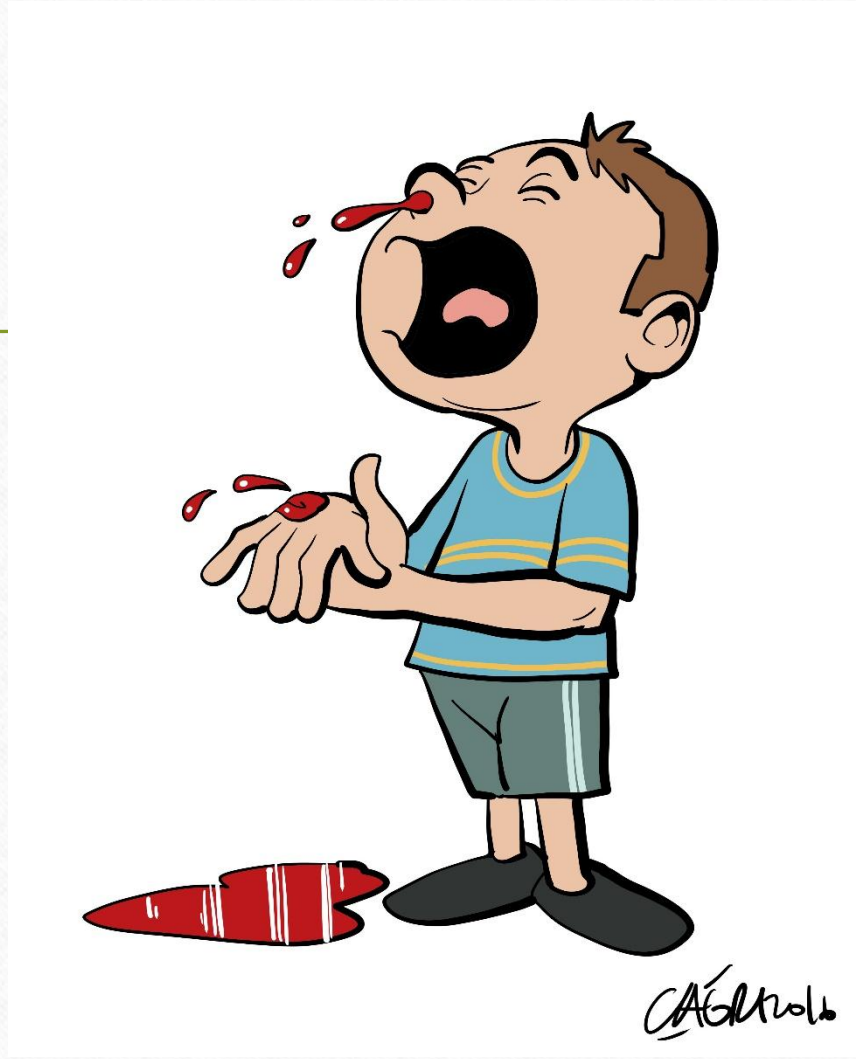
THE QUESTION

Whether a child has been abused, or

they might have a bleeding diathesis ?



- An incorrect diagnosis of child abuse can be devastating for the family and,
- If a serious underlying blood disorder is later identified, regaining the trust of the family may be very difficult.

- When a child presents with bruising or bleeding, the main differential diagnoses are;

- Physiological
- Accidental bleeding,
- Non-accidental injury, or
- A bleeding diathesis.

- It is difficult to interpret because of;

- Not enough information was available; for example, incomplete histories had been taken or investigations were incomplete.

- This inevitably led to delays in confirming the cause of the bleeding and

- Meant that; if parents or carers contested a diagnosis of abuse, excluding a bleeding disorder was extremely difficult.

Presenting complaint

- How the injury occurred ?
- Assessment made as to whether this explains the injuries or bleeding seen ?
- Guidance is given that; abuse should be suspected, if there is significant bruising or bleeding with no history of trauma or a history inconsistent with the severity of the injury.
- However, fingertip bruising can be seen in children with a bleeding diathesis from normal physical interaction. A drug history should also be elicited, particularly one of anticoagulant use.

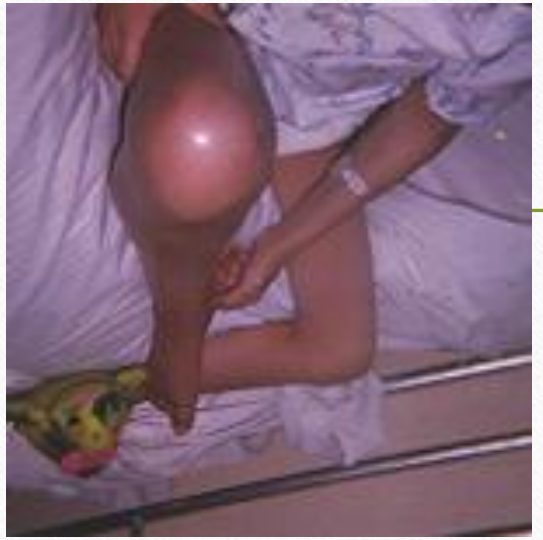
Significant haemostatic challenges

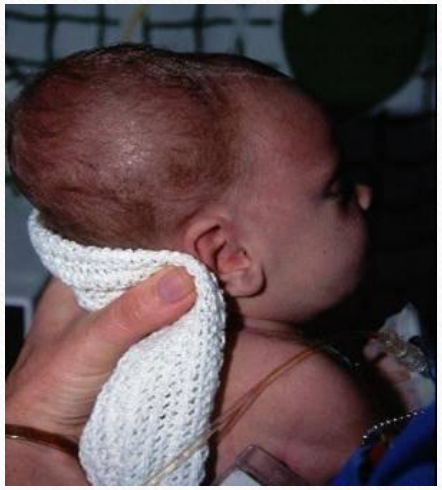
- Significant haemostatic challenges will include operations such as;
- circumcision,
- tonsillectomy,
- or removal of teeth.
- Bleeding response to injury such as a bitten tongue or a wound that requires stitching can yield useful information.

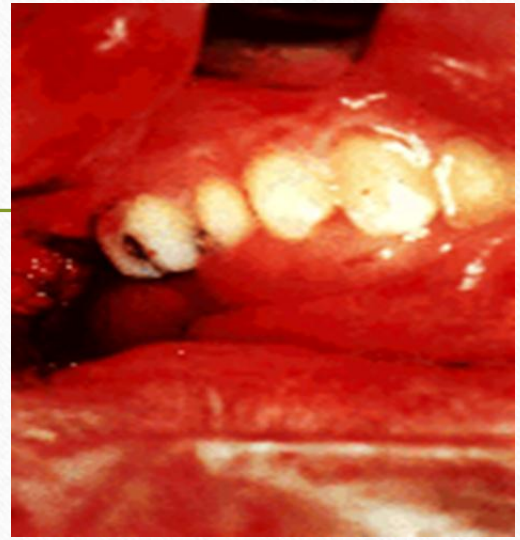
Significant haemostatic challenges

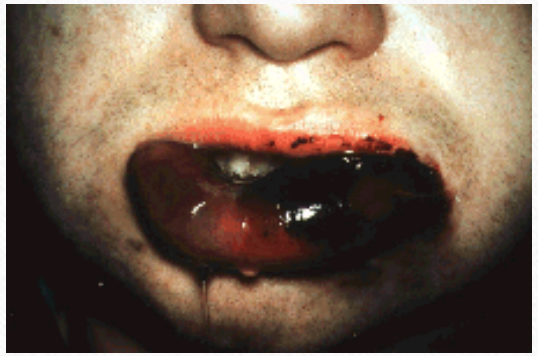
- Bleeding may be due to other disease states, that affect haemostasis such as hepatocellular dysfunction, renal disease, or malabsorption.
- In girls, menstrual loss may be an indicator of a bleeding diathesis.

Clinical Severity









Family history

- A family history of bleeding may be apparent and is more often seen in dominantly inherited or X linked conditions such as haemophilia A or B.

Examination of the child

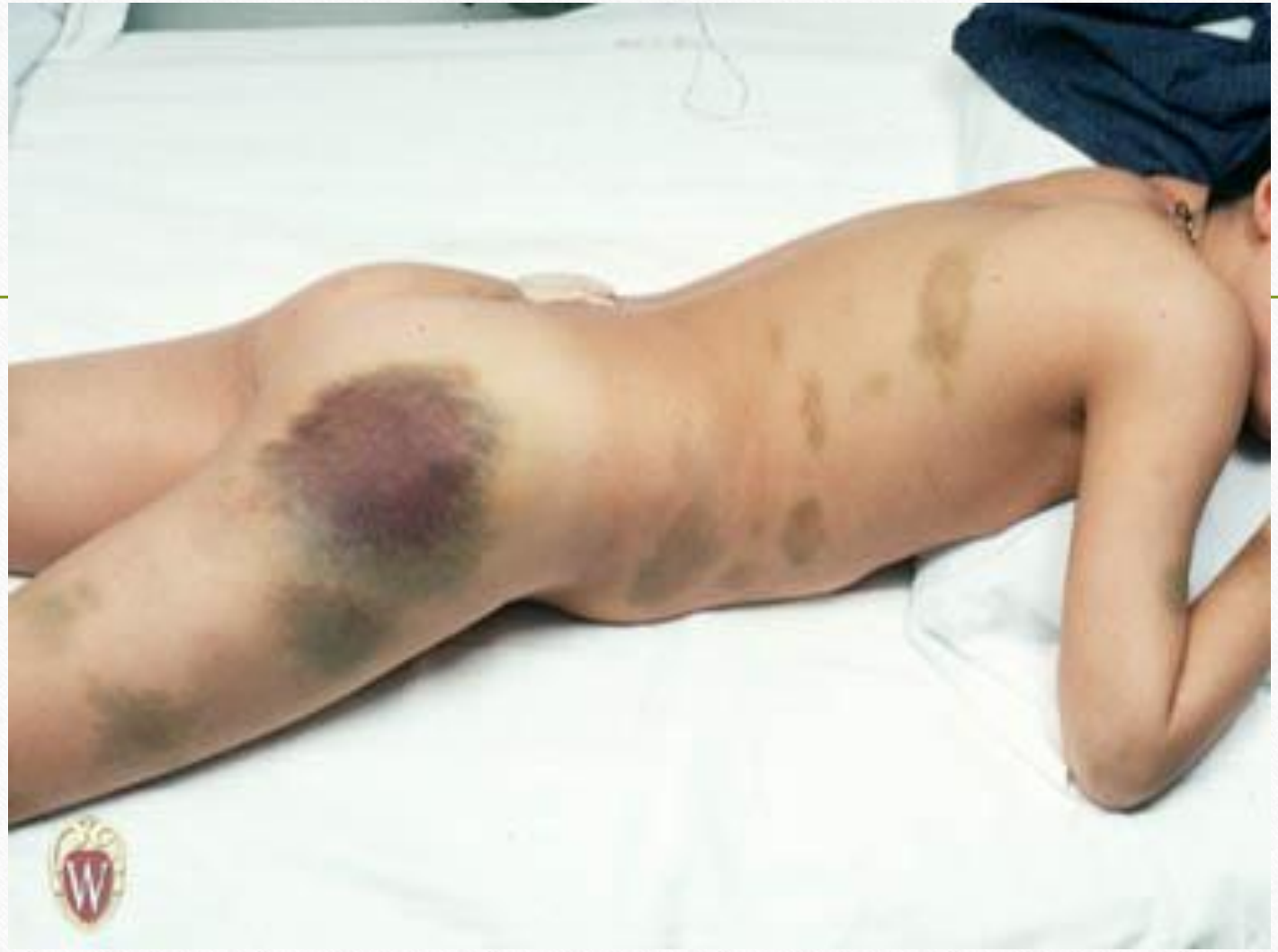
- When examining the child, the general health and state of the child should be assessed.

Bruises

- If the child is presenting with bruising, particular note should be made of the distribution and size of the bruises.
- Bruises of different ages are seen both in abuse and in children with a bleeding diathesis.
- The pattern of bruising should be recorded, in particular if marks indicate use of an object such as a belt or flex.

Bruises

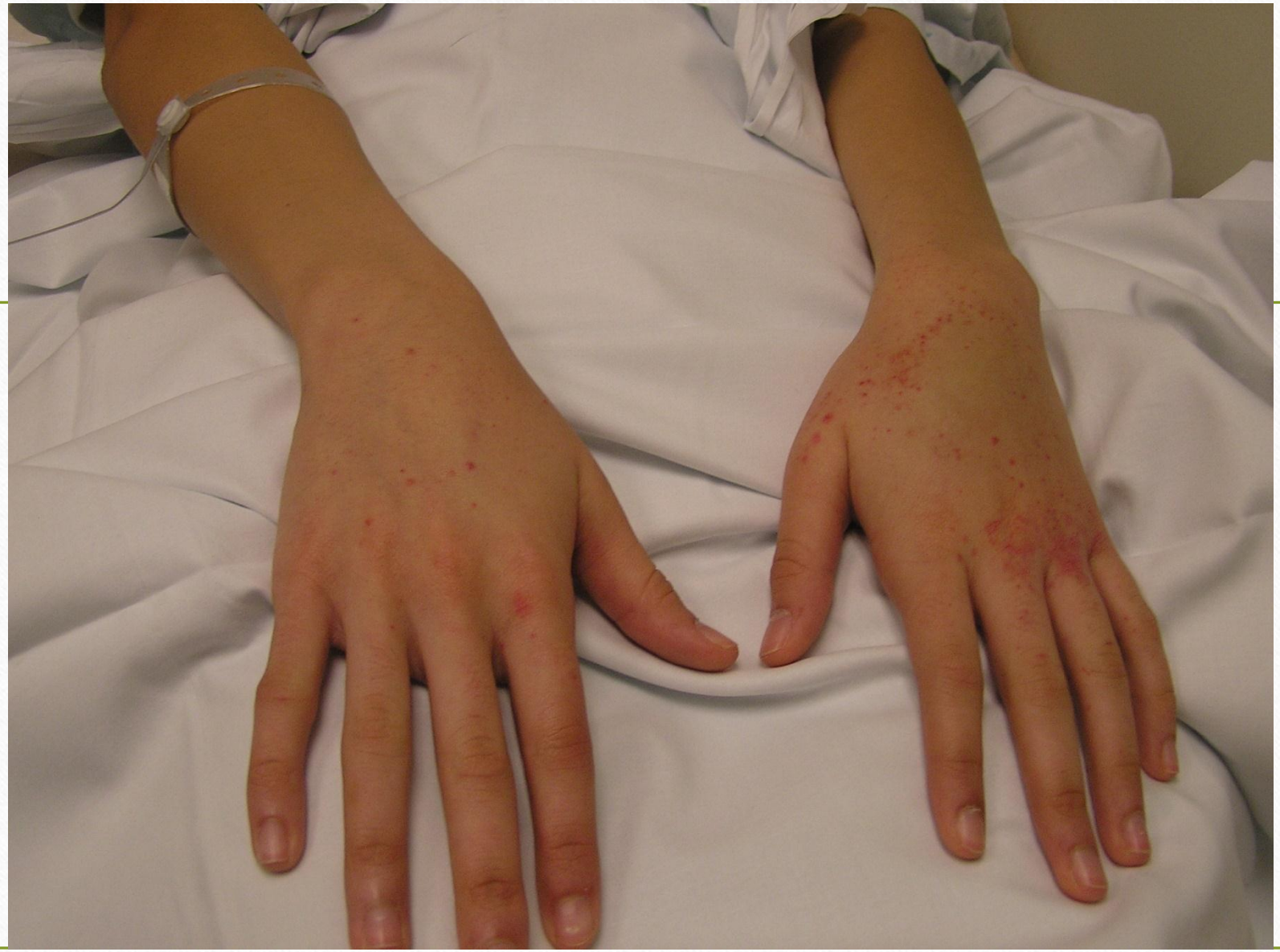
- The age of a bruise is very difficult to ascertain with certainty and depends on the integrity of the coagulation system and vessels and the force and location of the injury.
- Accompanying tissue swelling and abrasion may be present in more recent bruises.



Petechial haemorrhages

- Presence or absence of petechial haemorrhages will help differentiate disorders associated with thrombocytopenia.

- However such haemorrhages can occur in the distribution of the superior vena cava in association with a severe bout of coughing or vomiting in children without a bleeding diathesis or in cases of strangulation.



Bleeding into joints

- A swollen, tender joint may indicate a bleed into that joint as is seen in haemophilia,
- But tender joints may also be seen in Henoch-Schonlein purpura, acute leukaemia, or neuroblastoma.

Haematological investigation

- In a child who may have been abused, it is essential that the investigations are as atraumatic as possible and yield the maximum information.

First line investigations

- Initial tests should include a coagulation screen consisting of a prothrombin time (PT),
- activated partial thromboplastin time (aPTT),
- fibrinogen, and thrombin time (TT),
- plus a full blood count, platelet count, and blood film.

First line investigations

- A factor VIII, factor IX, and von Willebrand factor antigen and activity (Ristocetin cofactor) are also recommended in all cases of suspected non-accidental injury as a normal or

- Marginally prolonged aPTT can be associated with a significant decrease in factor VIII or IX levels or with von Willebrand disease.

First line investigations

- It is important to remember that some significant bleeding disorders give normal screen results. Such as;

- von Willebrand disease
- Henoch-Schonlein purpura
- Ehlers-Danlos
- Vitamin C deficiency

Platelet storage pool disorder

a2 antiplasmin deficiency

Glanzmann's thrombasthenia

First line investigations

- The pattern of abnormalities obtained using first line tests along with the clinical presentation and history may well indicate or identify any underlying disorder.

Test	Mechanism Tested	Normal Values	Disorder
Prothrombin time	Extrinsic and common pathway	<12 sec beyond neonate; 12-18 sec in term neonate	Defect in vitamin K-dependent factors; hemorrhagic disease of newborn, malabsorption, liver disease, DIC, oral anticoagulants, ingestion of rat poison
Activated partial thromboplastin time	Intrinsic and common pathway	25-40 sec beyond neonate; 70 sec in term neonate	Hemophilia; von Willebrand disease, heparin; DIC; deficient factors XII and XI; lupus anticoagulant
Thrombin time	Fibrinogen to fibrin conversion	10-15 sec beyond neonate; 12-17 sec in term neonate	Fibrin split products, DIC, hypofibrinogenemia, heparin, uremia
Bleeding time	Hemostasis, capillary and platelet function	3-7 min beyond neonate	Platelet dysfunction, thrombocytopenia, von Willebrand disease, aspirin
Platelet count	Platelet number	150,000-450,000/mm^3	Thrombocytopenia differential diagnosis
Blood smear	Platelet number and size; RBC morphology	-	Large platelets suggest peripheral destruction; fragmented, bizarre RBC morphology suggests microangiopathic processx(e.g., hemolytic uremic syndrome, hemangioma, DIC)

SPECIFIC DISORDERS
Inherited disorders

- Von Willebrand disease:
- The commonest of the inherited bleeding disorders with a prevalence of 1–2%
- Typical presentation is one of mucocutaneous bleeding.
- Inherited as an autosomal dominant
- Modestly reduced von Willebrand factor (vWF) levels that are associated with mild bleeding

Haemophilia

- Haemophilia A (factor VIII deficiency) and haemophilia B (factor IX deficiency), although rare, are the commonest inherited factor deficiencies associated with a bleeding diathesis.
- Both X-linked conditions and the severe forms occur almost exclusively in males.
- They are the commonest inherited bleeding disorders to present in the neonatal period.
- 90% of those with severe disease will have presented by the age of 1 year.

Haemophilia

- A coagulation screen in haemophilia A or B will show an isolated prolongation of the aPTT
- Haemophilia C
- Factor XI deficiency (haemophilia C) is a rare inherited bleeding disorder and is found mostly, but not exclusively among the Ashkenazi Jewish population.
- It is inherited in an autosomal recessive manner and bleeding is usually mucocutaneous in nature.
- A coagulation screen shows an isolated, prolonged aPTT.

Congenital platelet disorders

- The majority of the inherited platelet disorders that are associated with bleeding result in a degree of thrombocytopenia;
- Bernard Soulier syndrome,
- Wiskott Aldrich syndrome,
- May Hegglin anomaly,
- Glanzmann's thrombasthenia.

Congenital platelet disorders

- Glanzmann's thrombasthenia;
- A severe condition where the patient's platelets lack the IIb-IIIa receptor essential for binding of fibrinogen and platelet aggregation.
- There is a positive history of bleeding from birth, with mucocutaneous bleeding, spontaneous bruising, and significant bleeding with minor trauma.
- It is inherited as an autosomal recessive condition and thus there is usually no family history. Diagnosis is made from platelet function tests.

Acquired disorders

- Reduced vitamin K dependent factors
- Can occur in the neonatal period or early infancy.
- Early presentation may be secondary to maternal ingestion of vitamin K antagonists such as warfarin or anticonvulsants, or to lack of vitamin K prophylaxis at birth.
- Later presentation is commonly associated with exclusive breast feeding and either lack of vitamin K at birth or a single oral dose only.

Reduced vitamin K dependent factors

- About 50% of babies present with intracranial haemorrhage with high morbidity and mortality.
- Misdiagnosis of haemorrhagic disease of the newborn as child abuse has been reported.
- The coagulation screen shows a prolonged PT, with a variably prolonged aPTT dependent on the severity of the deficiency.

Thrombocytopenia

- Easily identified from the full blood count.
- The most likely cause is idiopathic thrombocytopenic purpura (ITP),
- The haemoglobin and white cell count is usually normal as is the coagulation screen.

Thrombocytopenia

- Other causes of acquired thrombocytopenia;
- Bone marrow infiltration from malignant disease or leukaemia,
- Disseminated intravascular coagulation are associated with additional abnormalities of the blood count or coagulation screen, and are seen in an ill child.

Approach to bleeding child: Summary

- Identify any specific defect of hemostasis
- Congenital vs. acquired / primary vs. secondary
- Screening investigation for bleeding disorder

Remember !!!

- It is important to remember that diagnosis of a bleeding diathesis, especially if associated with a mild phenotype, does not exclude non-accidental injury, and where these are found concurrently, the child will be at even greater risk.

Before everything, stop bleeding !!!!

- THANKS FOR YOUR ATTENTION