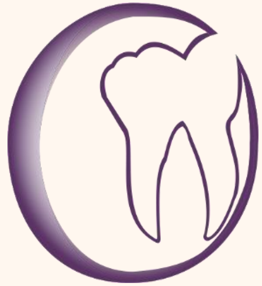
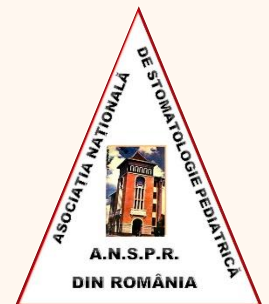


the Child Dental Patient with

Hereditary clotting disorders
- Haemophilia -



Haemophilia

- Definition: Hereditary clotting disorders due to factor VIII, IX or XI deficit (*).

(* Merck Manual, 1999)

HAEMOPHILIA

A

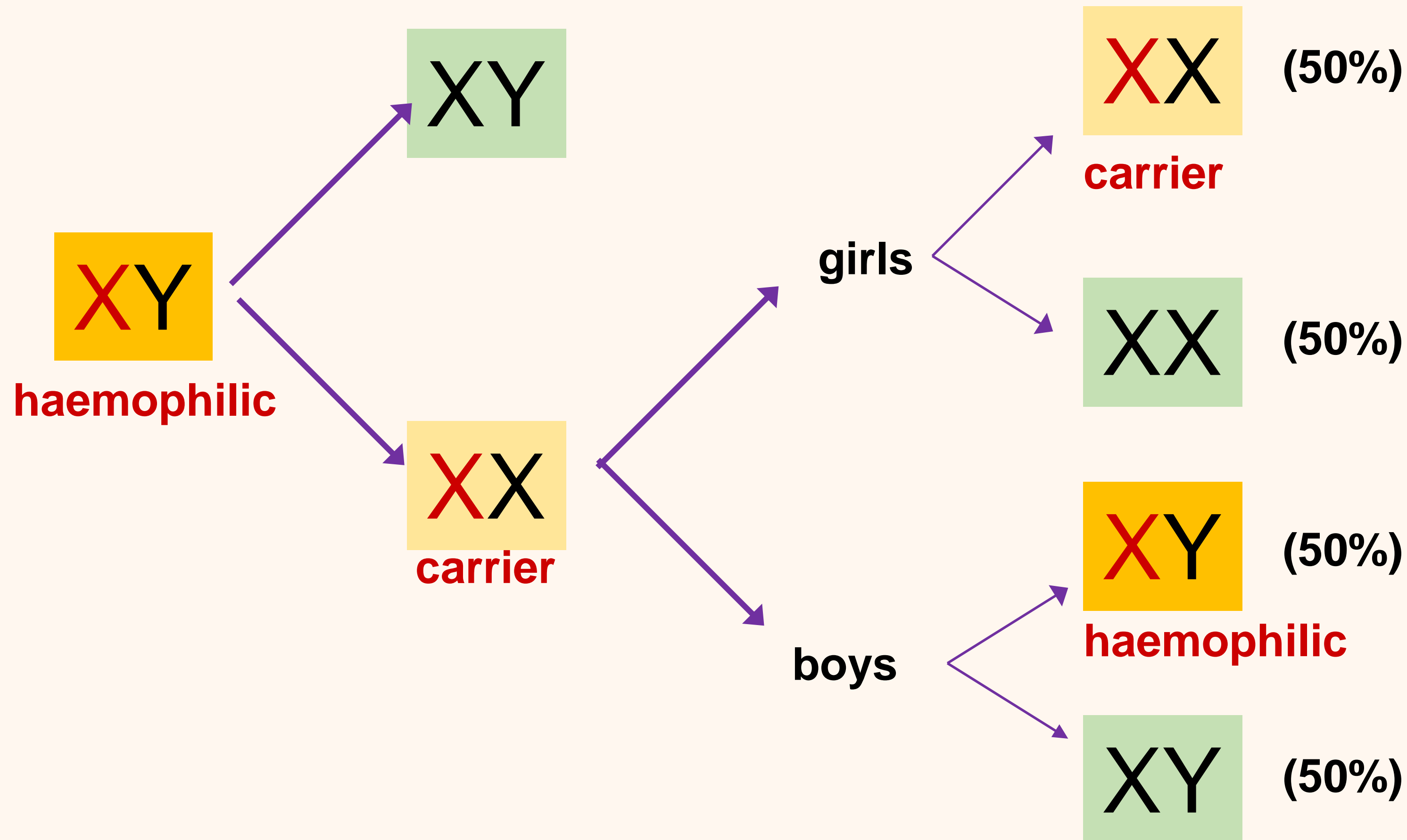
- clotting factor VIII deficit
- 80% of all haemophilias
- 1: 10.000 live male births

B

- clotting factor IX deficit
- "Christmas" Disease

- hereditary X-linked recessive transmission
- similar clinical features – diff. dg. - by specific tests (e.g. TGT)
- normal bleeding time, increased clotting time and Howell time

Genetic transmission of haemophilia



Females can also have haemophilia, but much rarer - when both X chromosomes are affected/one is affected and the other is missing or non-functioning. Bleeding symptoms can be similar to males with haemophilia.

Haemophilia

severe

- < 1% clotting factor
- profound spontaneous bleeding in muscles and joints, cerebral bleeding

moderate

- 2-5% clotting factor
- milder bleedings, usually after minor trauma/ bleeding therapeutic procedures

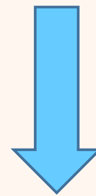
mild

- 5-40% clotting factor
- longer bleeding after important trauma/ extractions

Dentistry ⟷ Hereditary clotting disorders

The first sign in very mild clotting disorders is heavy bleeding from a dental procedure, an accident, or surgery.

(Merck Manual, 1999)



The dentist can be the first medical professional in the position to suspect the disorder

Factors to consider in the management of a hemophiliac dental patient (*)

- Dental neglect → need for frequent extractions
- Trauma and surgery
- Factor VIII inhibitors
- Hazards of anesthesia and injections
- Risk of hepatitis B,C and liver disease and HIV infection
- Aggravation of bleeding by drugs
- Anxiety
- Drug dependence

(*)Scully C. Medical problems in dentistry. 6th Ed. Elsevier, London 2010.

Dental treatments in haemophiliac children

⇨ Endodontic therapy - non vital techniques are preffered – do not require protection

⇨ Infiltration anesthesia - only under protection

⇨ Extractions - under haemathological control

- require protection
- hospitalization

Restorative dental treatment

- avoid brusque maneuvers during dental treatment
- prevent accidental damage to the oral mucosa (saliva ejectors, placement of X-ray films/ sensors)
- atention when using matrix bands, rubber dam, wooden edges→ risk of bleeding
- tests and precautionary measures - HIV, hepatitis B, C.

Preventive treatment

- Toothbrushing twice/day using medium texture brush
- Fluoride containing toothpaste:
  - 1000 ppm for children under 6 years old
  - 1400+ ppm for children over 6 years old.
- Dental floss or interdental brushes when possible
- Regular dental visits - starting with the first tooth eruption, every 6 months
- Early interception of orthodontic problems – avoid need for fixed appliances if possible (risk of bleeding), minimize crowding (can favour plaque retention and gingivitis, with increased risk of bleeding)

Dental anesthesia

- local infiltration and periodontal ligament injection are preferred
- nerve-block injections (inferior alveolar and posterior superior alveolar) can cause airway obstruction
- anesthetic block or intramuscular injections must be preceded by replacement therapy in all cases
- if FVIII levels<50% of the normal reference value: anesthetic block or intramuscular injections are contraindicated.

Extractions and dentoalveolar surgery

- All necessary surgery - in one operation (if possible)
- Factor VIII level of 50–75% is required
- Maintenance of normal Factor VIII levels for approx one week – in major surgery procedures and for moderate and severe patients
- Non-traumatic needle, minimum number of sutures and resorbable sutures are recommended
- Post-operatively, a diet of cold liquid and minced solids - for to 5–10 days

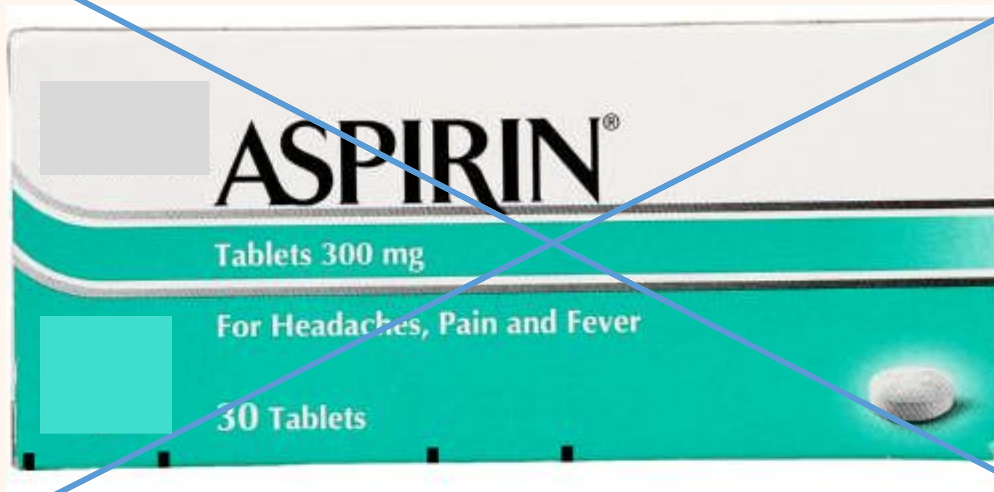
Dental care under GA (*)

- Intubation can cause submucosal hemorrhages (can be life threatening).
- Avoid nasal intubation - can be traumatic; bleeding from the site can lead to aspiration.
- Carefully positioning of the extremities and pressure points padding - to prevent intramuscular hematomas or hemarthrosis.

* Rayen R et al. (2011). Dental management of hemophiliac child under general anesthesia. Journal of the Indian Society of Pedodontics and Preventive Dentistry 2011; 29 74-9.

Pain control

- Aspirin and it derivatives must be avoided
- Acetaminophen and paracetamol = safe alternatives



For haemophilic children are mandatory:

- very good oral hygiene – to avoid gingivitis and gum bleeding
- regular and frequent dental check-ups
    - to avoid complicated caries lesions
    - interceptive therapy
- strict measures of asepsis and antisepsis - avoiding the spread of infectious diseases
- parents' awareness about the consequences of the disease

Possible questions from parents

- tooth **eruption** - normal

(!) traumatizing the overlaying gingiva during eruption may cause bleeding

- deciduous teeth **shedding** - normal

(!) extended mobility may impose extraction under protection

- **orthodontic** treatment - yes, under haematological control.

Interception if possible