Women's Issues in Bleeding Disorders
Menorrhagia, pregnancy and delivery

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Women's Issues

- Menorrhagia in women with bleeding disorders
- Inheritance and genetic counselling
- Symptomatic haemophilia carriers
- Pregnancy and delivery
- Some issues for an affected baby

Women Bleed Too

http://www.womenbleedtoo.org.uk

(UK Haemophilia Society)

www.projectredflag.org

(NHF of USA)

‘Females with von Willebrand disease:
72 years as the silent majority’

(PA Kouides, Haemophilia 1998; 4: 665-676)

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Menorrhagia can be defined objectively or subjectively

Objectively, menorrhagia is taken to be a total menstrual blood loss = or >80 ml per menstruation

Subjectively, menorrhagia is defined as a complaint of excessive menstrual blood loss occurring over several consecutive cycles in a woman of reproductive years

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What is menorrhagia?

- Menstrual flow that soaks through one or more sanitary pads or tampons every hour for several consecutive hours
- The need to use double sanitary protection to control menstrual flow
- Soaking through bed clothes
- Menstrual period that lasts longer than 7 days
- Menstrual flow that includes large blood clots
- Associated with low iron stores (ferritin) and anaemia

Menorrhagia in women with bleeding disorders

Disorder	Prevalence of menorrhagia %
Von Willebrand Disease	74-92
Bernard Soulier syndrome	51
Glanzmann thrombasthenia	98
Factor XI deficiency	59
Carriers of haemophilia	57
Other rare factor deficiencies	35-70

(Reproductive health in women with bleeding disorders Kadir R and James AH – WFH Monograph 2009)

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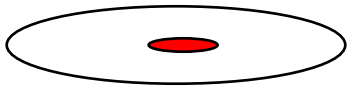
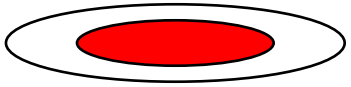
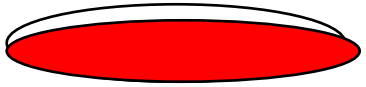
PICTORIAL ASSESSMENT OF MENSTRUAL BLOOD FLOW

Days of your period

	1	2	3	4	5	6	7	8
Pad								
1 pt 								
5 pts 								
10 pts 								
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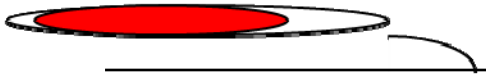
5 pts 								
10 pts 								

Record for each day the number of pads or tampons which match each illustration

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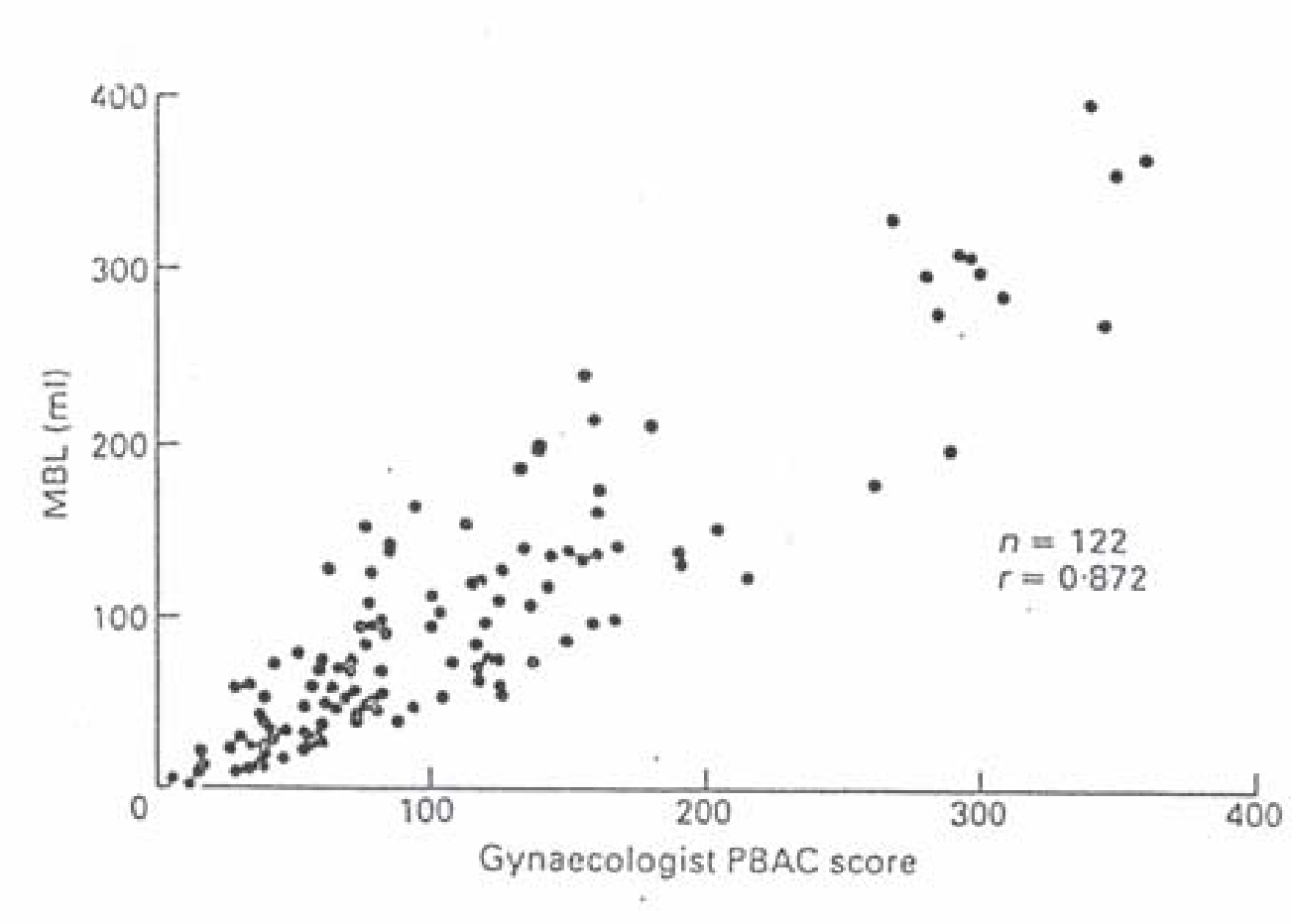
PBAC scores against menstrual blood loss



Pictorial Blood Assessment Chart
score 100 = 80ml blood

Higham et al BJOG 1990; 97: 734-9

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Women with inherited bleeding disorders

- 116 women studied at Royal Free Hospital
  - 66 vWd
  - 30 carriers of haemophilia
  - 20 with factor XI deficiency
- Menstrual loss assessed with the PBAC
  - Menorrhagia defined as score >100
- Age matched control group of 69 women

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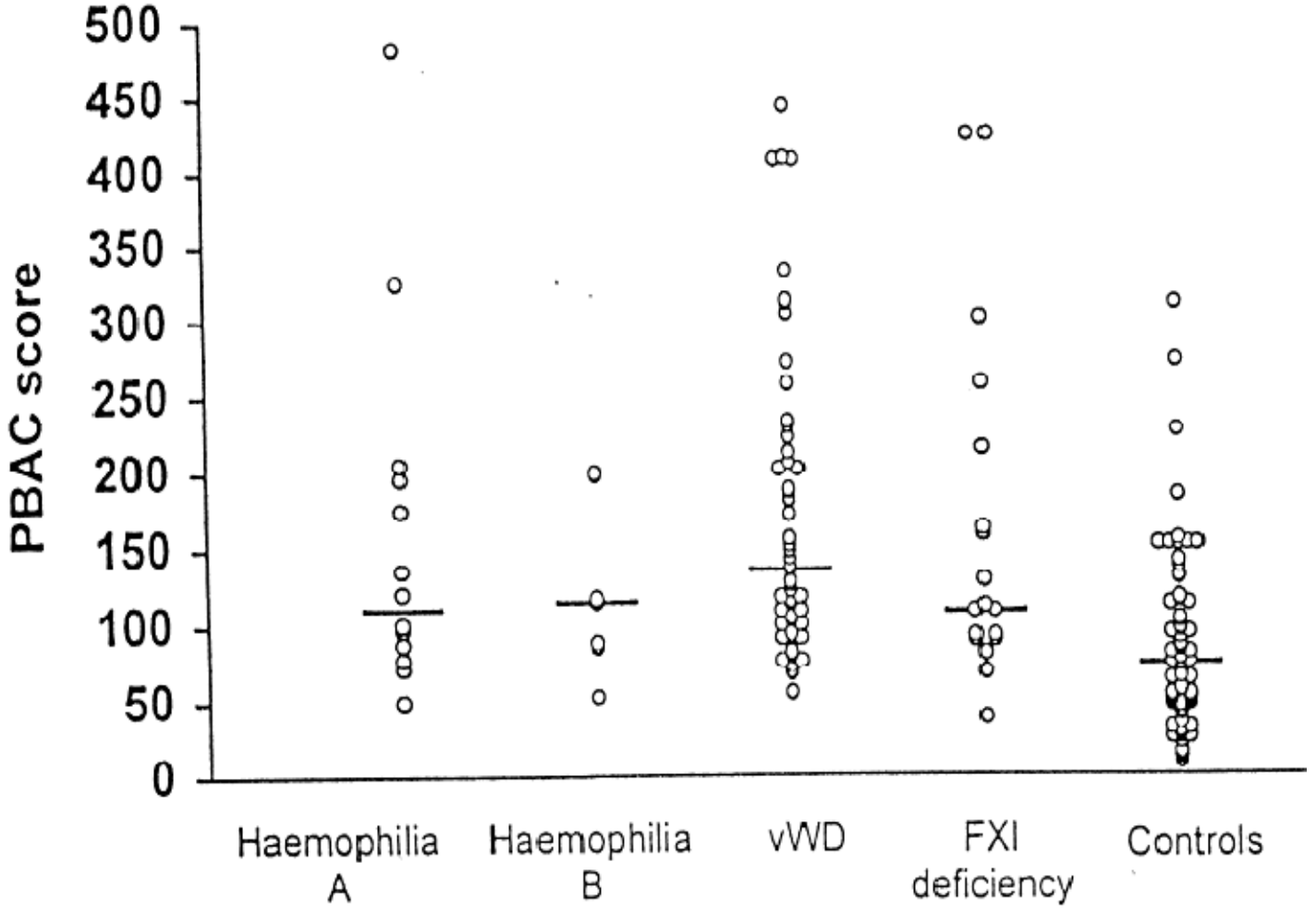
Kadir et al. Haemophilia 1999; 5: 40-48

Menstrual scores in women with bleeding disorders compared with controls



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Kadir et al. Haemophilia 1999; 5: 40-48

Frequency of inherited bleeding disorders in women with menorrhagia

- 12% general gynaecology referrals are for menorrhagia
- 208 women attending gynaecology clinic RFH October 1995 – June 1997 screened with bleeding history including PBAC
- 150 women with PBAC score > 100 –

  APTT; FVIIIC; VWF AC; and FXIC

Kadir et al Lancet 1998; 351: 485-89

Frequency of inherited bleeding disorders in women with menorrhagia

Number	150
Mild VWD	15
Moderate VWD	3
Factor XI deficiency	4
Combined deficiencies	2
Carrier of haemophilia A	1
Platelet disorder	1

Kadir et al. Lancet 1998; 351: 485-489

Frequency of von Willebrand disease in women with menorrhagia

- 150 women attending gynaecology clinic with PBAC score of >100
  - 15 Mild VWD
  - 3 Moderate VWD
- Frequency of VWD 13% compared with maybe 0.1 to 1% of the normal population

Kadir et al. Lancet 1998; 351: 485-489

VWD in women: a systematic review

- 11 studies including 988 women
- 131 diagnosed with VWD
- Prevalence mean 13% (5-24%)
- Testing for VWD should be included in the investigation of menorrhagia

Shankar et al BJOG 2004; 111: 734-40

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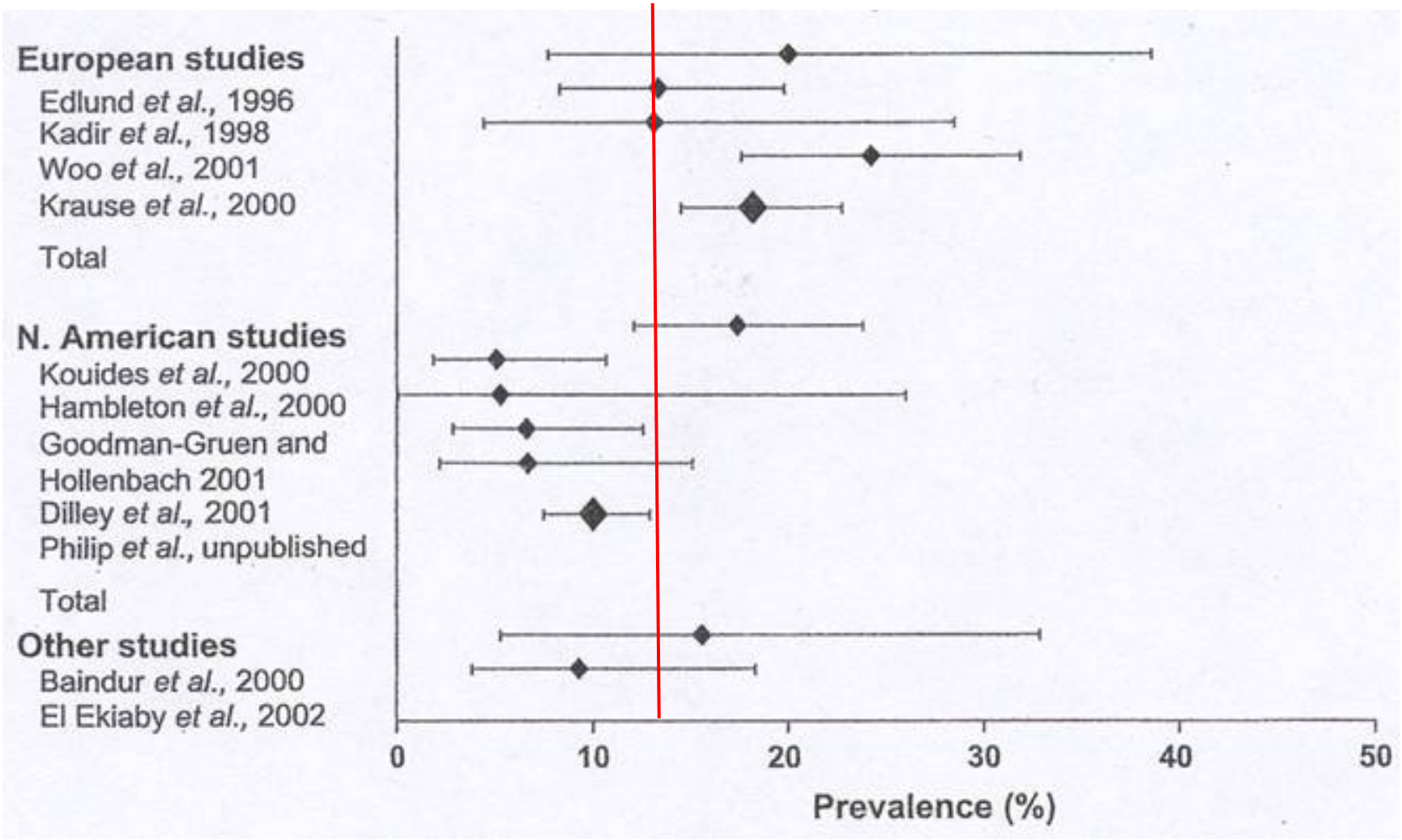
Prevalence rates of von Willebrand disease in 988 women presenting with menorrhagia



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Shankar et al BJOG 2004; 111: 734-40

The frequency of bleeding symptoms in the normal population compared with 264 Scandinavian patients with VWD

(Silwer, 1973; Mauser Bunschoten et al, 1988; Nosek-Cenkowska et al, 1991; Sramek et al, 1995)

Investigated symptom	Frequency	
	normal	VWD
Epistaxis	5-39%	62%
Bruising	12-24%	49%
Bleeding from small wounds	0.2-2%	36%
Gum bleeding	7-51%	35%
Menorrhagia	23-44%	60%
Post partum haemorrhage	6-23%	23%
Bleeding after tooth extraction	1-13%	51%
Bleeding post tonsillectomy	2.4-11%	
Bleeding after surgery	1.4-6%	28%
AT LEAST ONE SYMPTOM		
MALE	>25%	
FEMALE	>46%	

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An underlying bleeding disorder should be suspected when a woman with menorrhagia has:

- Family history of a bleeding disorder
- Personal history of
  - Nose bleeds >10 mins
  - Notable bruising without injury (>2cm)
  - Minor wound bleeding (cuts >5 mins)
  - Excessive bleeding after dental extractions/surgery
  - Bleeding from ovarian cysts or corpus luteum
  - PPH especially delayed >24h

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Menorrhagia and quality of life

- Studied in 99 patients with inherited bleeding disorder (vWD 57, haemophilia carriers 24, and FXI deficiency 18) compared with 69 normal controls
- Menorrhagia
  - Prolonged duration
  - Flooding
  - Passage of clots
- Associated with reduced quality of life
  - Interference with work
  - Pain

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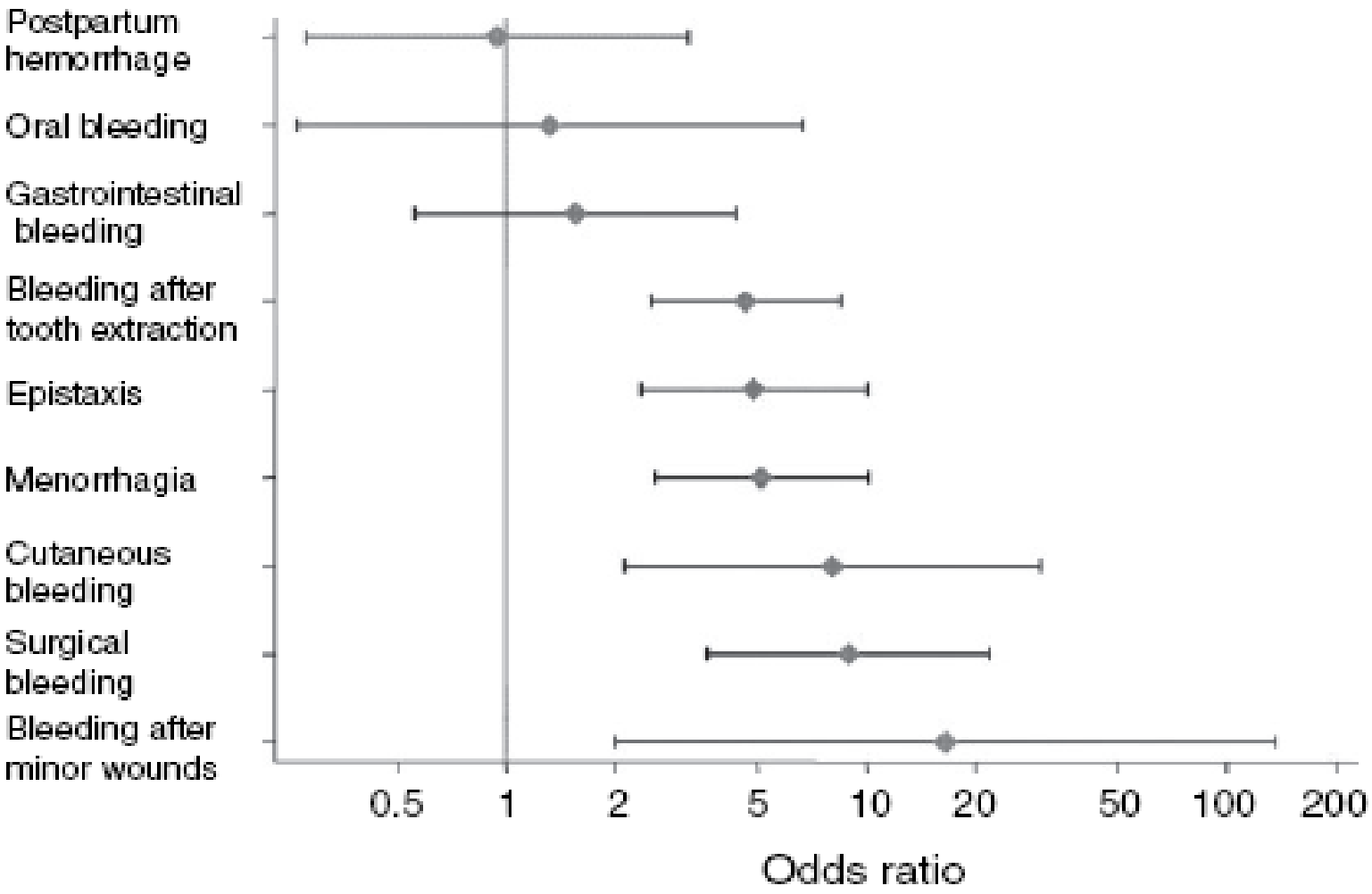
Kadir et al Haemophilia 1998; 4: 836-841

Predictive value of bleeding symptoms in diagnosis of type 1 VWD (Tosetto et al JTH 2006)



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Treatment of Menorrhagia

- Oral iron
- Tranexamic acid 1 g three or four times a day
- Combined oral contraceptive or similar.
- Norethisterone 5-10 mg three times daily for 5 days before or during period (or continuously) (titrate dose)
- Intranasal or subcutaneous desmopressin on day 1 and 3 of period
- Factor replacement therapy
- Levonorgestrel intra-uterine system (Mirena coil)
- Injectable medroxyprogesterone acetate
- If future fertility not required, then endometrial ablation or hysterectomy

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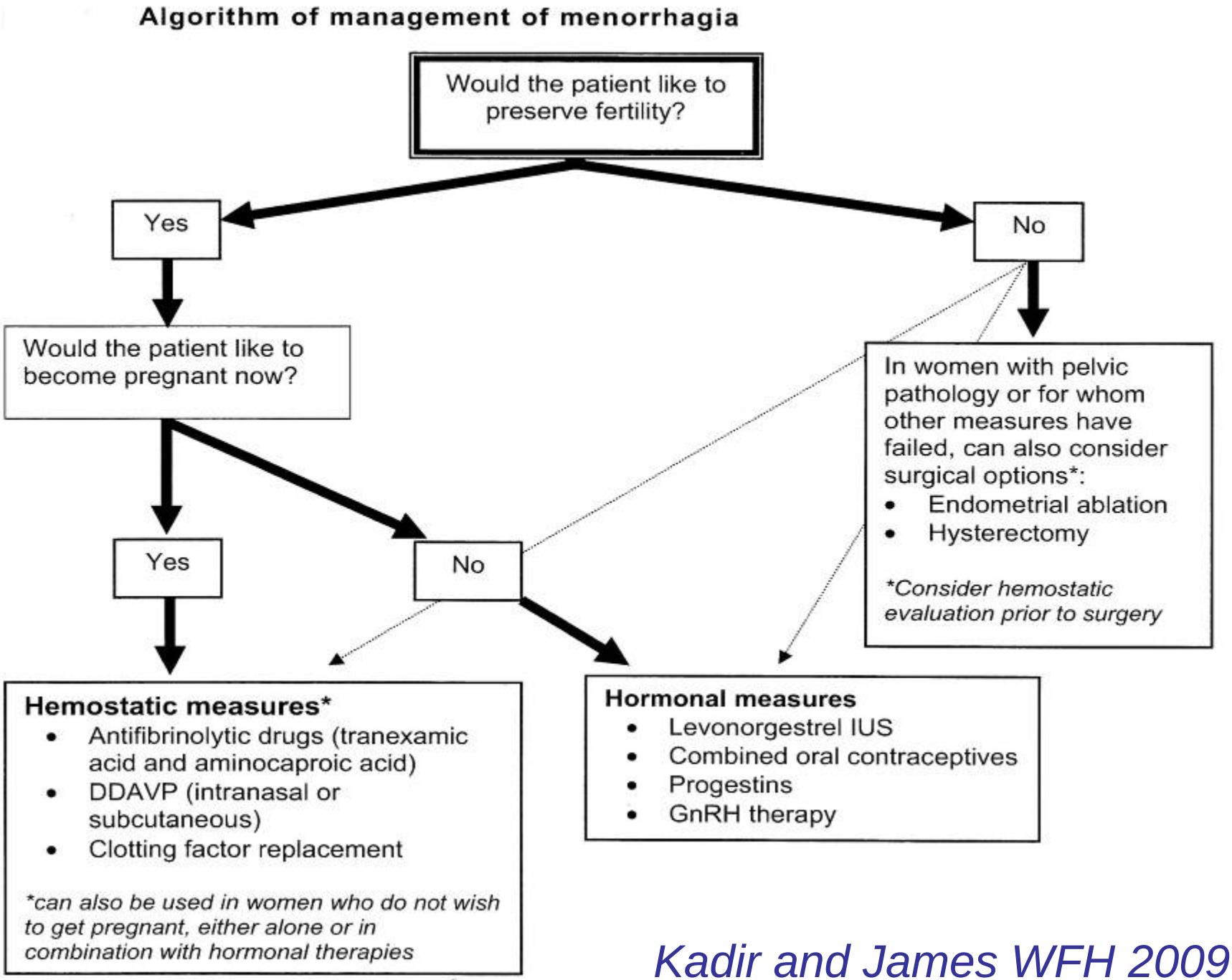
James et al. Consensus for management of VWD and other bleeding disorders in women experiencing menorrhagia or PPH. Am J Obs Gyn 2009; 20:12 e1-8



Kadir and James WFH 2009

Haemorrhagic ovarian cysts

- Multiple case reports (9/136, 7% women with vWD in one series)
- Can be severe
- Bleeding into peritoneal cavity or retroperitoneum
- Correct the bleeding disorder
- Ovulation is not normally accompanied by any clinically significant bleeding
- Increased risk of endometriosis

Review of pregnancy and childbirth in vWd

- 4067 deliveries in women with vWd between 2000-2003 (USA)
- Diagnosis of vWd 1 in 4000 women
- Increased risk of antepartum bleeding odds ratio 10.2
- Increased risk of PPH odds ratio 1.5
- Higher risk of transfusion odds ratio 4.7
- 5 maternal deaths, a rate 10 fold higher than normal women but rate still very low – 1 in 1000 women

James A and Jamison MG, JTH 2007; 5: 1165-1169

Rare Bleeding Disorders

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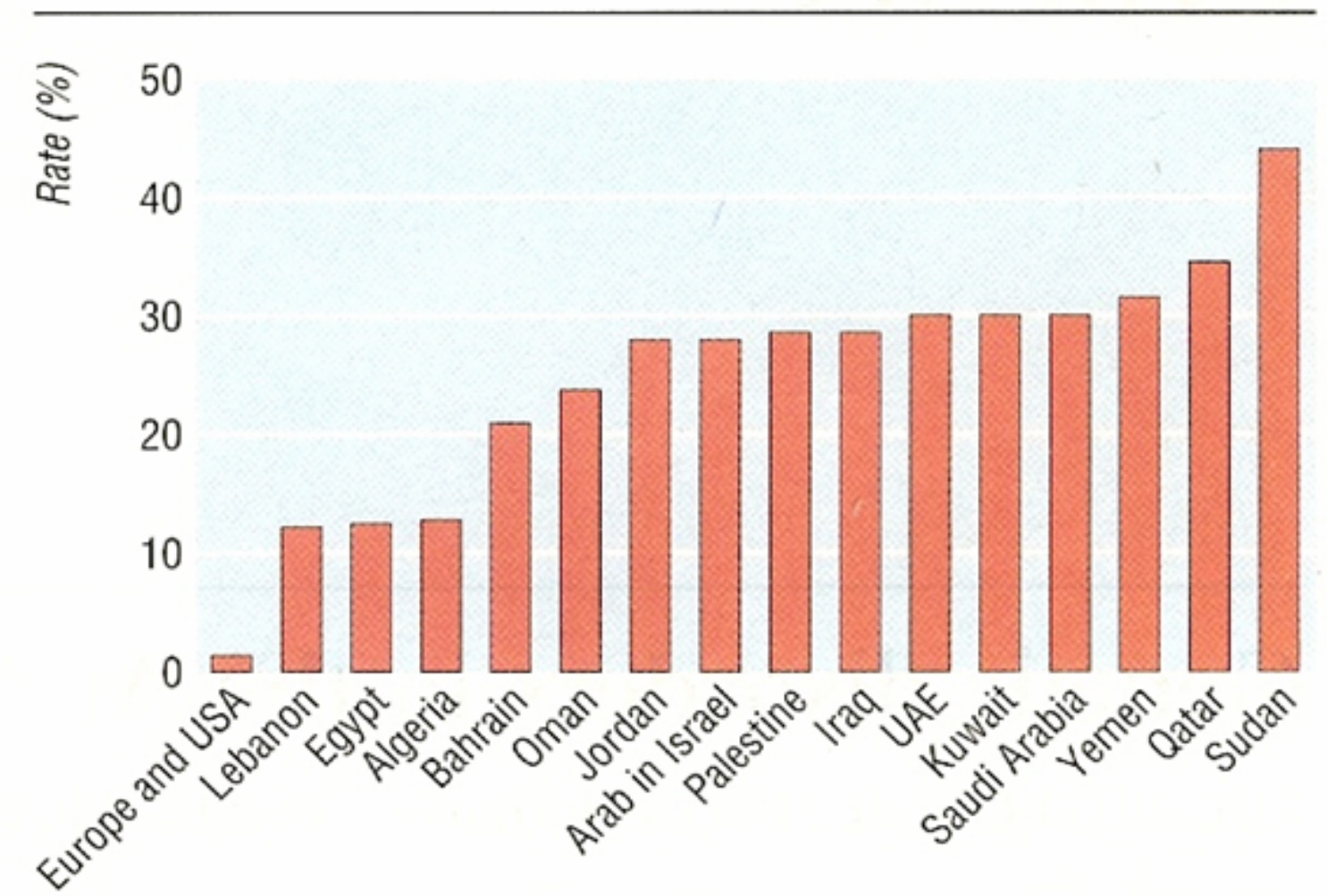
Average rates of marriages between first cousins among Arabs



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Al-Gazali L. et al BMJ; 333: 831-4 J 2006

Gynaecological and obstetric problems in women with different bleeding disorders

- **A comparison of 114 women with bleeding disorders and 114 normal women**
  - 48 vWd
  - 31 haemophilia carriers
    - 15 with normal levels
    - 15 mild deficiency
    - 1 moderate deficiency
  - 35 rare bleeding disorders
    - 14 FVII
    - 10 FXI
    - 4 V+VIII
    - 7 others (fibrinogen, XIII, V and X)
  - 10 severe (<1%)
  - 8 moderate (2-10%)
  - 17 mild (11 to LLN)

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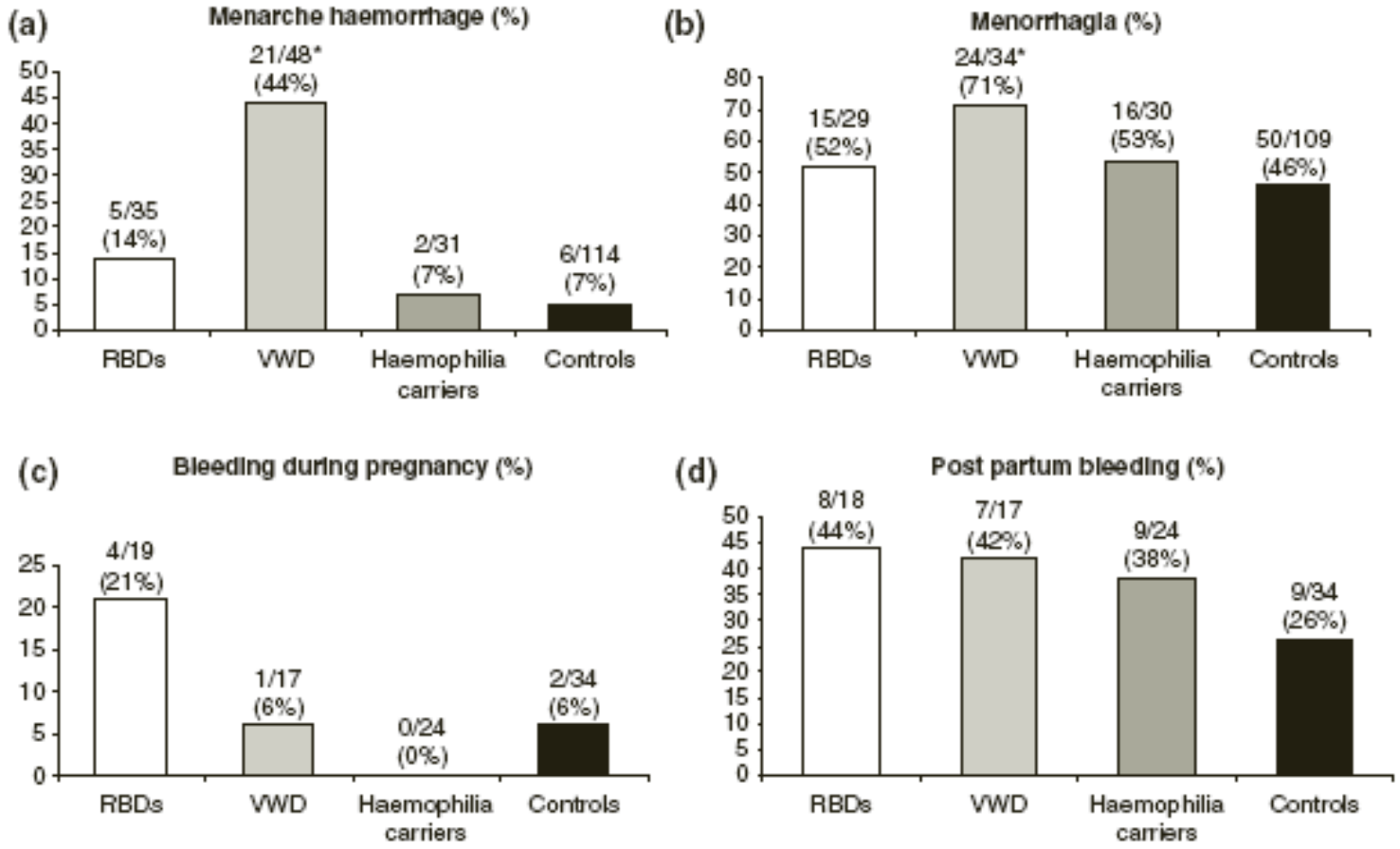
Siboni et al. Haemophilia 2009 (online June 2009)



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Siboni et al. Haemophilia 2009

Factor VII deficiency and menstruation

- 14 women with FVII deficiency (2 severe)
- 23 healthy women
- PBAC and quality of life questionnaire
- Menorrhagia = PBAC score >100
  - FVII deficient 8/14 (57%) and 6 (43%) anaemia
  - healthy women 4/23 (17%) 2 (9%) anaemia
- Significantly worse quality of life scores in women with FVII deficiency

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Kulkarni et al. Haemophilia 2006; 12: 248-252

Adolescents

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Acute menorrhagia in adolescents

- 9-year case review January 1971-80
- Admissions to hospital excluding genital tract pathology
- Primary coagulation disorder
    - 20% of total 59 patients
    - 25% of those with HB < 10g/dl
    - 33% of those requiring transfusion
    - 50% of those presenting at the menarche

Claessons and Cowell Am J Obstet Gynecol 1981; 139: 277-80

Menorrhagia in adolescents

- Adolescents < 20 years admitted to University of Michigan hospital 1979-95.
- 37 adolescents with 46 admissions.
- Menarche, average 12.9y.
- Admission, average 15.9y.
- 15/46 (33%) haematological disease.

Smith et al J Pediatr Adolesc Gynecol 1998; 11: 13-5

Inheritance and genetic counselling in the bleeding disorders

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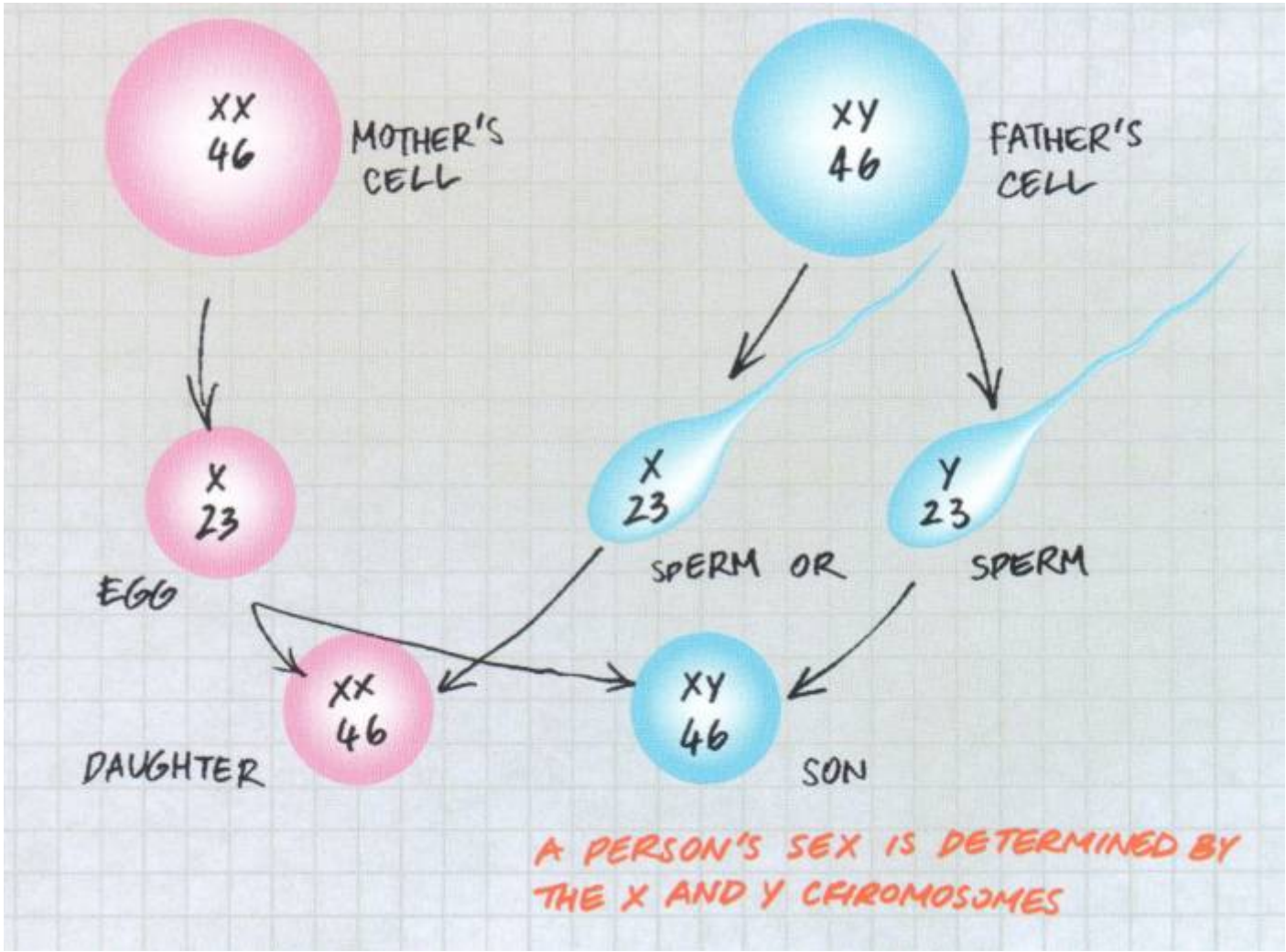

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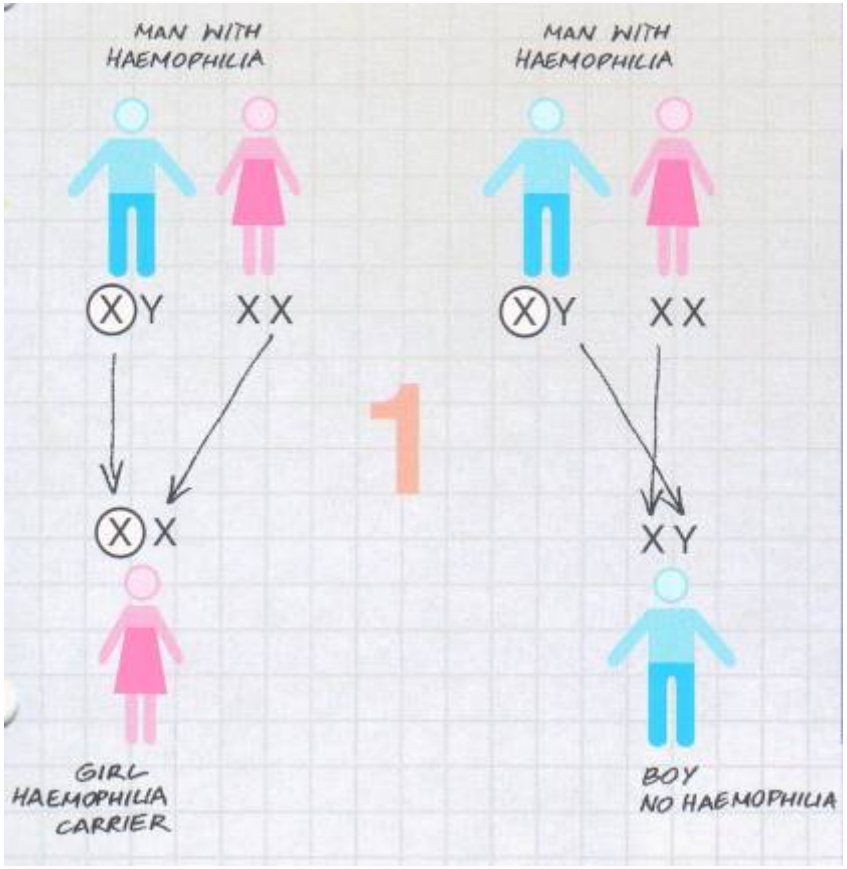

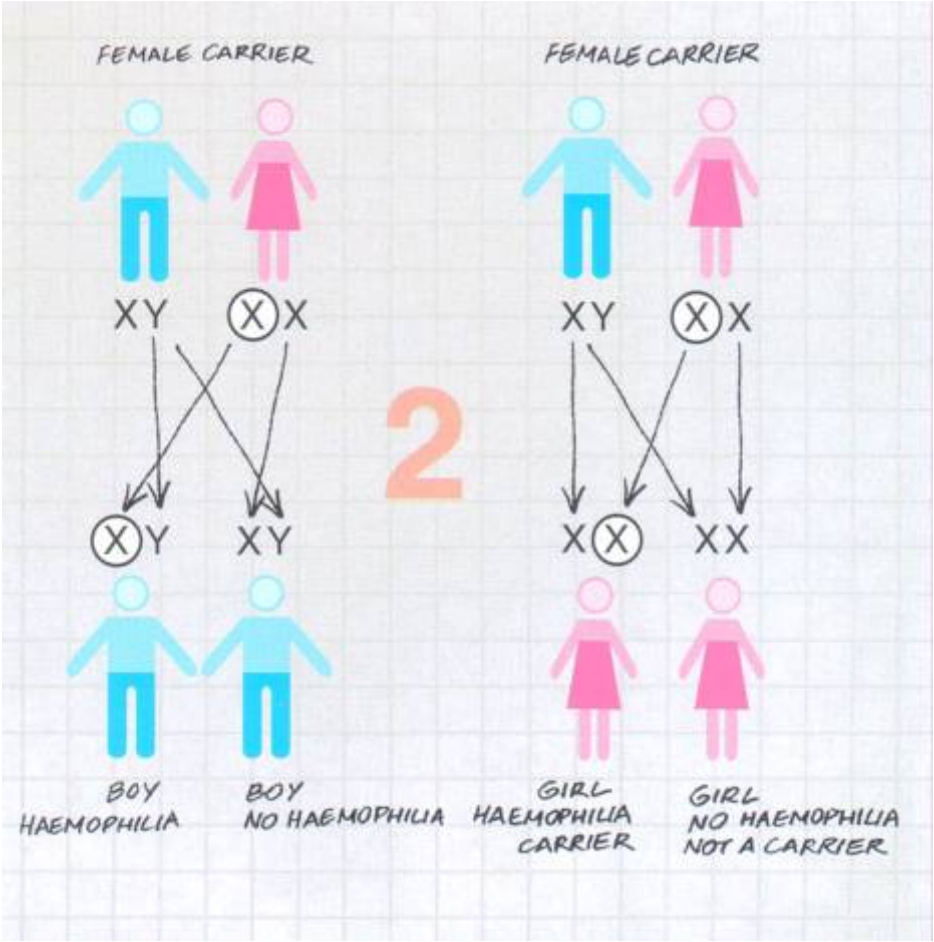

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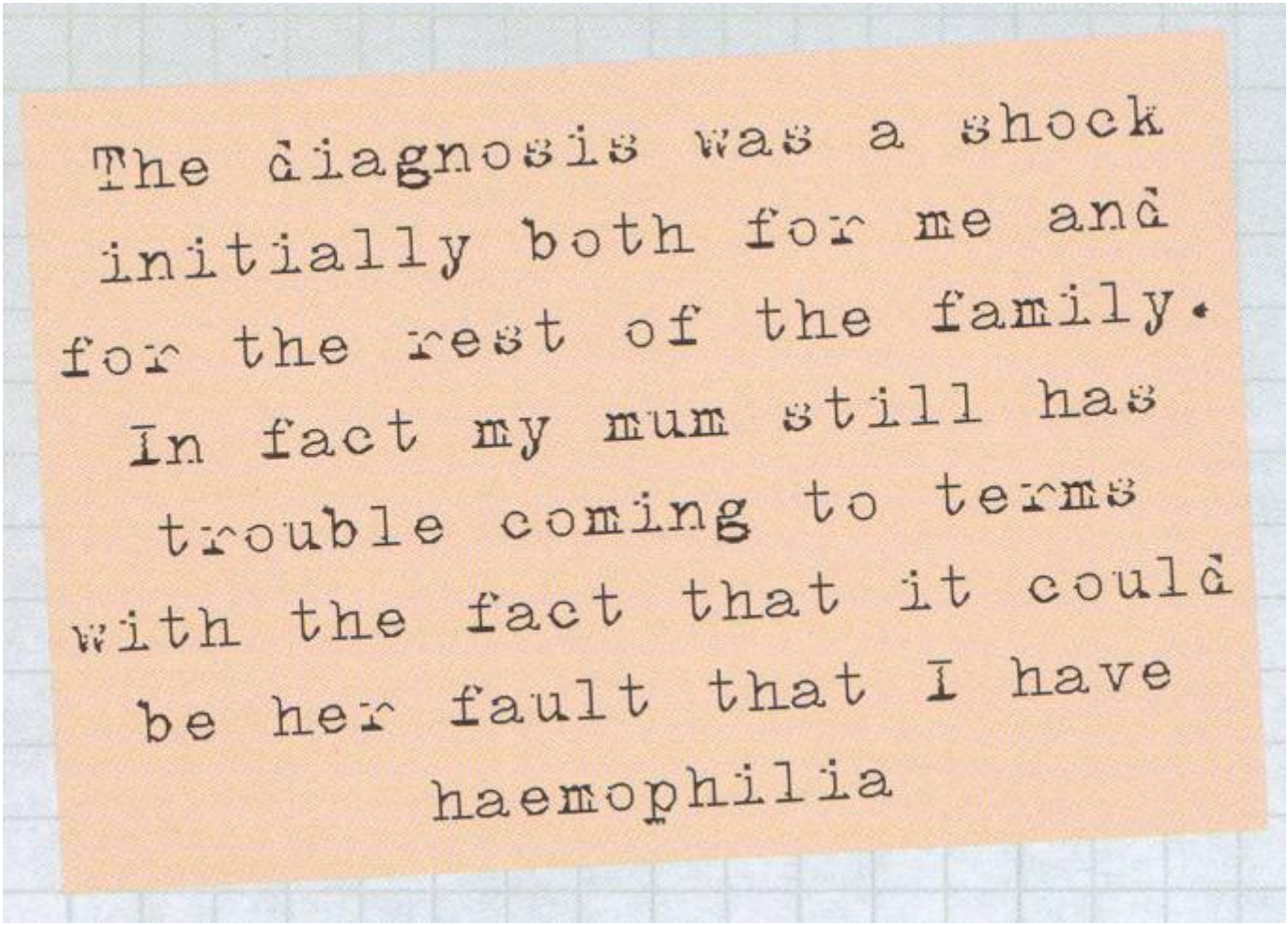

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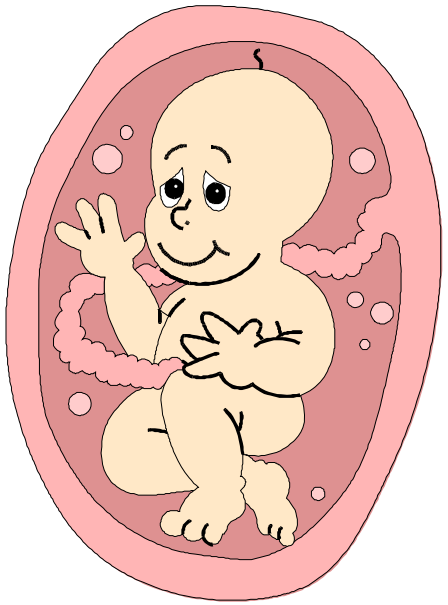
Congenital bleeding disorders

- Too often unprepared in a family with known bleeding disorder
- Screen family members
- Counselling about risks
- Antenatal diagnosis



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Haemophilia and inheritance

- Carriers of haemophilia A or B may be at risk of excessive bleeding and need levels measuring early in life
- A normal level does not exclude carrier status
- Implications of inheritance should ideally be discussed well in advance of pregnancy
- Education needs to begin early
- Reproductive choices

Information about carrier status

- 1990 a study of 549 obligate and potential carriers found that
  - 41% had never been tested and were not informed that they might be at risk
  - 19% of women had never discussed the inherited nature of the disease in the home
- Ideally:
  - Age 1 yr check FVIII or IX level
  - Teenage – undertake genetic testing
  - Adult woman and partner – pre-pregnancy counselling
- Problems with discovery of carrier status – stigmatisation, guilt

Management of Haemophilia A and B Carriers

- Average factor level in carriers is 60% (range 5 to 219) compared to 100% in normal women (range 45 to >250)
- 8% carriers HA have a level <50% at delivery
- 50% carriers HB have a level <50%
- Cut off for treatment of carriers for surgery or delivery:
  - >60 no bleeding so no interventions required
  - <60 be careful – there is evidence of excessive bleeding after surgery with levels 40-60%
  - <30 treat

Plug et al. Blood 2006; 108: 52-56

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Genetic counselling in the haemophilia centre-constraints

- It is essential that those seeking genetic counselling are free to make decisions that are not constrained by their commitment to existing family members.
- Haemophilia specialists are highly committed to the success of treatment and may or may not be conscious of the potential impact of this on their ability to remain neutral in the counselling situation.
- Can issues of paternity be discussed in the haemophilia centre where staff and patients know each other so well?

(Ludlam et al. Haemophilia 2005, 11: 145-163)

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Genetic testing – the process

- Informed consent with presentation of written material
- Provide the opportunity for questions
- Keep a copy of the signed consent form in the notes, and give one to the person
- Make clear the arrangements for delivery of results
- Ensure the information has been understood

What information?

- The potential clinical effects of being a carrier or affected person
- Current treatment and implications of the condition
- The mode of inheritance and the individual's genetic risk
- The rationale for identifying the genetic defect

Genetic counselling and genetic testing

- The rare bleeding disorders are autosomal recessive disorders
- Often the parents are consanguineous
- Parents should be advised of the risk of having further affected children
- Antenatal diagnosis and termination of pregnancy may be available

Challenges of Genetic Counseling in Developing Countries

- Inadequate diagnostic, management , rehabilitation facilities, burden of inherited disorders are great
- Social, culture, educational and religious background differs
- Availability of prenatal diagnosis

Data courtesy of Shirin Ravanbod, Iran

Why consanguineous marriage is favoured

- Easy acceptance of genetic disorder or carrier status in close relatives
- No religious and social division
- Family economic considerations

Data courtesy of Shirin Ravanbod, Iran

The Meaning of Carrier Status giving birth to unhealthy children

- Intense emotions;
  - Fear, anxiety, sadness, anger, guilt
- Stigma (discrimination around sisters)
- Society and in-laws pressure
- Marriage (vulnerable situation)
- Abortion

Slide courtesy of Shirin Ravanbod, Iran

Contacting relatives

- Generally it is the responsibility of the affected person to inform members of the family
- It is recommended that a post-consultation letter be sent indicating the genetic risks, options available and the offer of genetic counselling to other at-risk relatives

Genetic testing in children

- Males with haemophilia should have their genotype established
  - Inhibitor risk
  - Helpful to identify other affected family members
  - Must send written information about results

Genetic testing in children

- Females who are potential carriers
  - Assay factor because it might affect clinical management (low in a third of carriers)
  - Genetic testing deferred until girl can give informed consent (usually aged 12 years upwards)

Management of pregnancy and delivery

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Haemostasis in Pregnancy

Unchanged:

- **Factors II, V, IX, XIII**

Increased:

- **Fibrinogen (x2), factor VII, Factor VIII (x2), VWF (x3-4), factor X**

Decreased:

- **Factor XI (?)**
- **Platelets in 3rd trimester.**

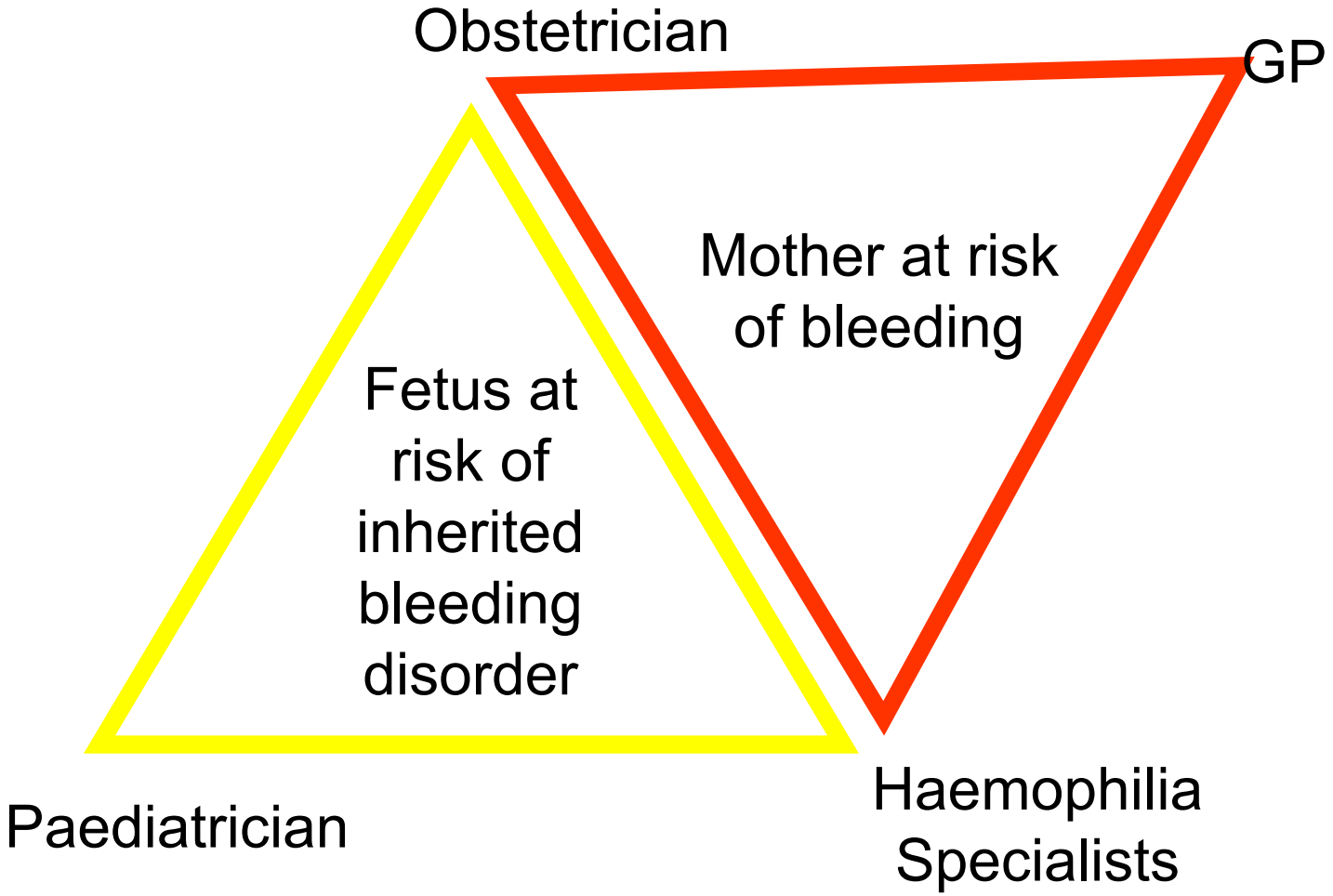
Reproductive choices

- Male fetus
  - Is he affected?
  - Termination or not
- Adoption
- Avoid children altogether
- Preimplantation genetic diagnosis
  - Selection of unaffected embryos
  - Many questions and concerns

Pregnancy and the Haemophilia Carrier

- Pre-pregnancy counselling and genetic analysis
- Fetal sexing by
  - Fetal DNA analysis from maternal blood at 8-9 wks
  - ultrasound in second trimester
- Antenatal diagnosis
  - Preimplantation genetic diagnosis – creation of embryos by IVF and selection of unaffected one
  - Chorionic villus sampling (1% risk of miscarriage), performed at 10.5 wks, termination by 13 wks
  - Ultrasound guided fetal blood sampling at 18-20 weeks for coagulation testing – greater risk of miscarriage, rarely performed now

Good communication between specialists is vital – joint clinics



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Management of pregnancy and delivery

- Monitor VIIIC in mid trimester.
- Formulate delivery plan in final trimester.
- Correct VIIIC at the beginning of labour.
- Umbilical sample at birth for urgent VIIIC.
- Affected males given prophylactic treatment with concentrate at birth?
- No IM injections (give vitamin K orally)

Pre-conceptual Counselling Joint Consultation

↓

Book early. Dating scan. Levels FVIII/FIX

↓

Fetal sexing: Fetal DNA from maternal blood at 8-9 week
PND - CVS between 11-13 weeks (if severe disease)
+/-Prophylaxis if FVIII or FIX < 50IU/dl

↓

Re-affirm fetal sex at Anomaly Scan. Review in ANC

↓

Review at 32 weeks.
Assess FVIII/FIX levels (if not normal at booking)

↓

Review at 36 weeks. Joint consultation

↓

Formulate ***Delivery Care Plan***
based on levels at 32 weeks.

↓

Aim for spontaneous vaginal delivery

↓

Prophylaxis if FVIII or FIX < 50IU/l
Desmopressin/VIII concentrate or FIX concentrate at start of labour

↓

Regional analgesia if FVIII/FIX > 50IU/dl

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Pregnancy and VWD:

- Most type I VWD corrects completely to normal values
- Type II and III do not correct.
- Type IIb get worse with progressive thrombocytopenia
- Correct haemostasis at the start of labour
- Monitor RiCof post-partum since VWF may decline quite rapidly
- Post-partum, type II and III may require replacement therapy, and type I may need Desmopressin

Pregnancy and bleeding disorders: General

- Planned delivery with a delivery plan.
- No ventouse or scalp electrodes.
- ? Forceps.
- Increased risk of operative delivery.
- Correct bleeding disorder in mother at the start of labour.
- Severe deficiencies may require correction for seven days post-partum.

Women at risk of excessive bleeding at delivery

- Management plan prepared in advance
  - Several copies including to the woman
- Joint management with haemophilia specialists (joint clinic)
- Correct the bleeding disorder prior to delivery
- Discuss constraints on epidural anaesthesia in advance
- Aim for natural labour to reduce risks of operative delivery
- Constraints due to risk of birth of affected baby

Obstetric pain relief in women with bleeding disorders

- Study of 80 pregnancies in 63 women
  - 19 FXI deficiency
  - 16 Haemophilia A carriers
  - 15 von Willebrand disease
  - 7 platelet disorders
  - 2 other
- 72 women seen in a joint clinic

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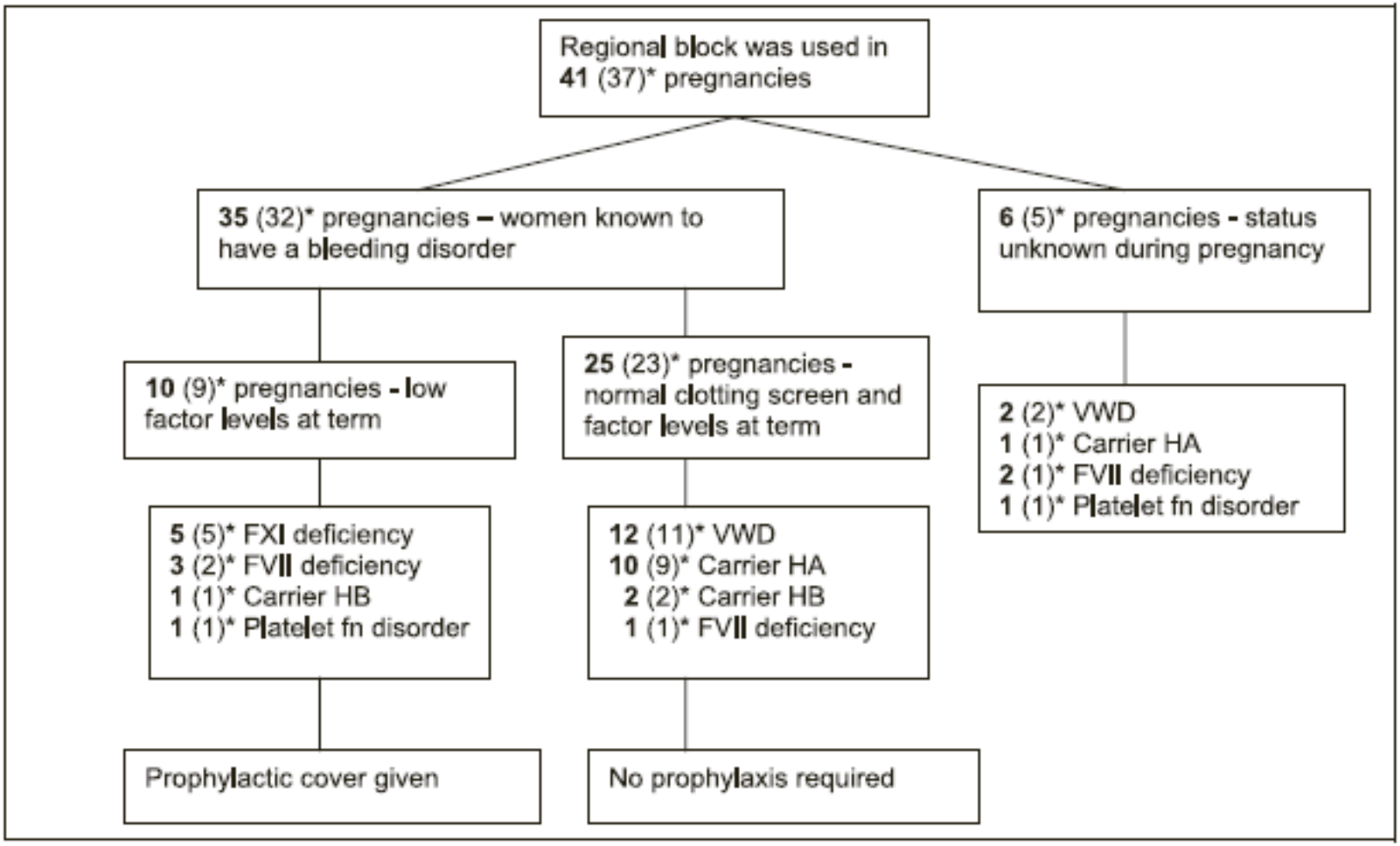
Chi et al. Thromb Haemostas 2009; 101: 1104-11

Obstetric pain relief in women with bleeding disorders

- Regional block performed in 41 pregnancies
  - 35 known to have a bleeding disorder
    - 10 given treatment
    - 25 not given because normal coagulation at term (vWD and HA carriers)
- FXI deficiency mostly managed with tranexamic acid
  - 4 with history of bleeding received concentrate
- Platelet disorders received platelet transfusions and TA at delivery

Use of regional block during labour and delivery



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Regional anaesthesia

- Advantages
  - Pain relief
  - Permits CS without GA
  - Earlier mobilisation
  - Earlier breastfeeding
  - Better for baby
- Disadvantages
  - Low blood pressure
  - Failure of block
  - Side effects including urinary retention
  - Bleeding with resultant paralysis (rare)

Regional anaesthesia at Royal Free Hospital

	All women	Bleeding disorder group
Regional anaesthesia	**45%**	**33%**
Caesarian sections under regional block	**93%**	**81%**

The complication rate was slightly higher in the bleeding disorder group

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Postpartum haemorrhage

- Primary – i.e. within first 24 hours
- Secondary – later, can occur after discharge from hospital.
  - Women with inherited bleeding disorders should be warned
  - Tranexamic acid is often helpful
- Von Willebrand levels drop rapidly after delivery, some within 6h

Management of the child with a bleeding disorder

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Risk of bleeding into the head in haemophilia

- Risk of intracranial bleeding in first 4 weeks about 4% haemophilic babies
- Normal babies 1 in 860 (vacuum extraction) to 1 in 1900 (normal deliveries)

Management of infants with haemophilia

- Prospective study of 580 babies diagnosed aged 0-2 yrs
- USA 135 haemophilia centres
- Birth and delivery – 68% vaginally, 32% CS
- Factor therapy at birth 45 (8%), 26 as prophylaxis (controversial) and 19 for bleeding events
- Head bleeding events as initial bleeding event
  - 28 ICH (71% in infants with no known family history)
    - 27 at less than 1 month of age, 17 of these associated with delivery
    - 5 events aged 1-6 months
- Bleeding after circumcision
- Bleeding after heel sticks

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Kulkarni et al. Haemophilia 2009

How can we identify the women at risk?

- **Target the women through advocacy organisations.**

- **Remind GPs and gynaecologists that menorrhagia is symptom of a bleeding disorder.**

- **Remind haematologists that women with a bleeding disorder may have menorrhagia.**

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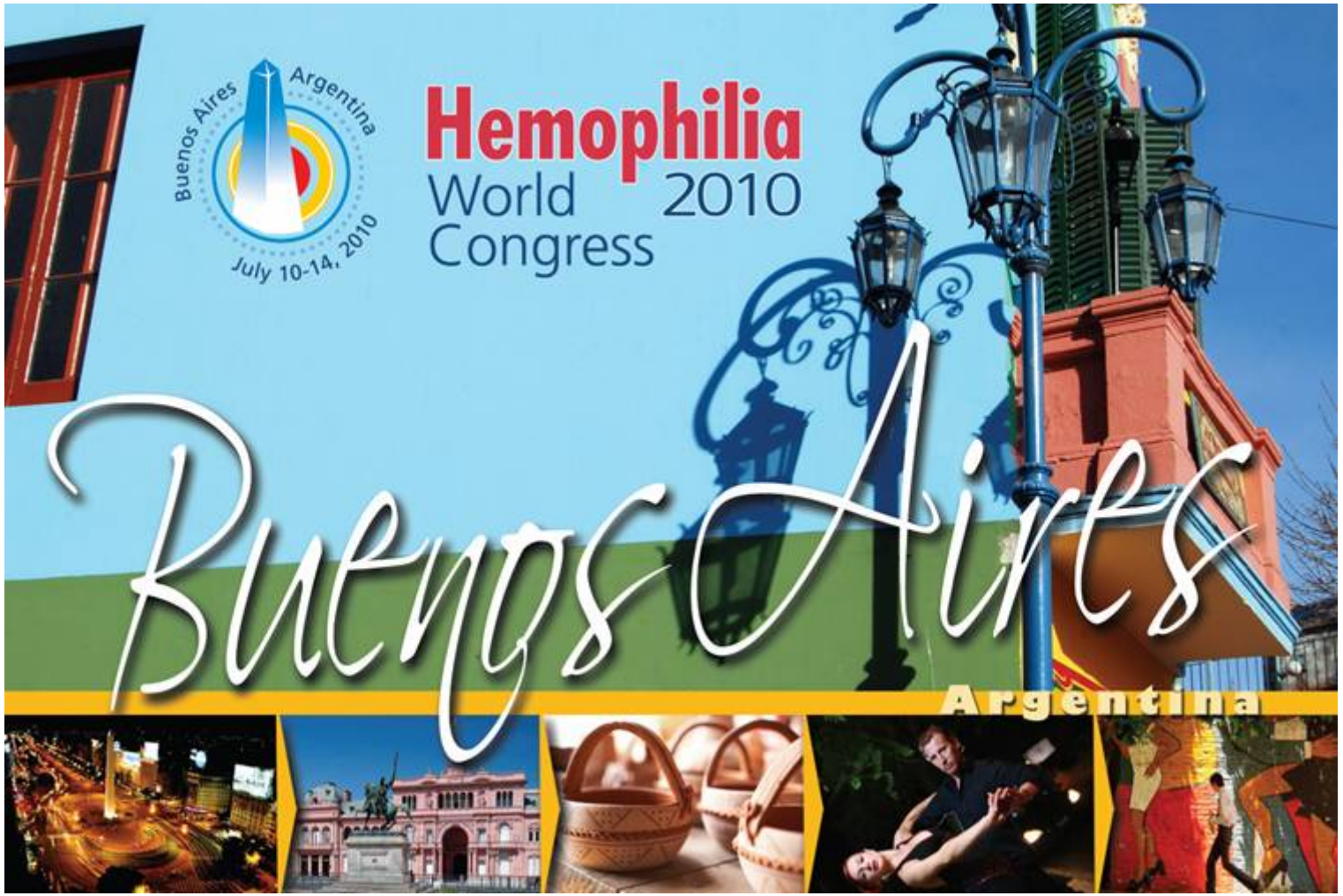
Hope to see you there



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