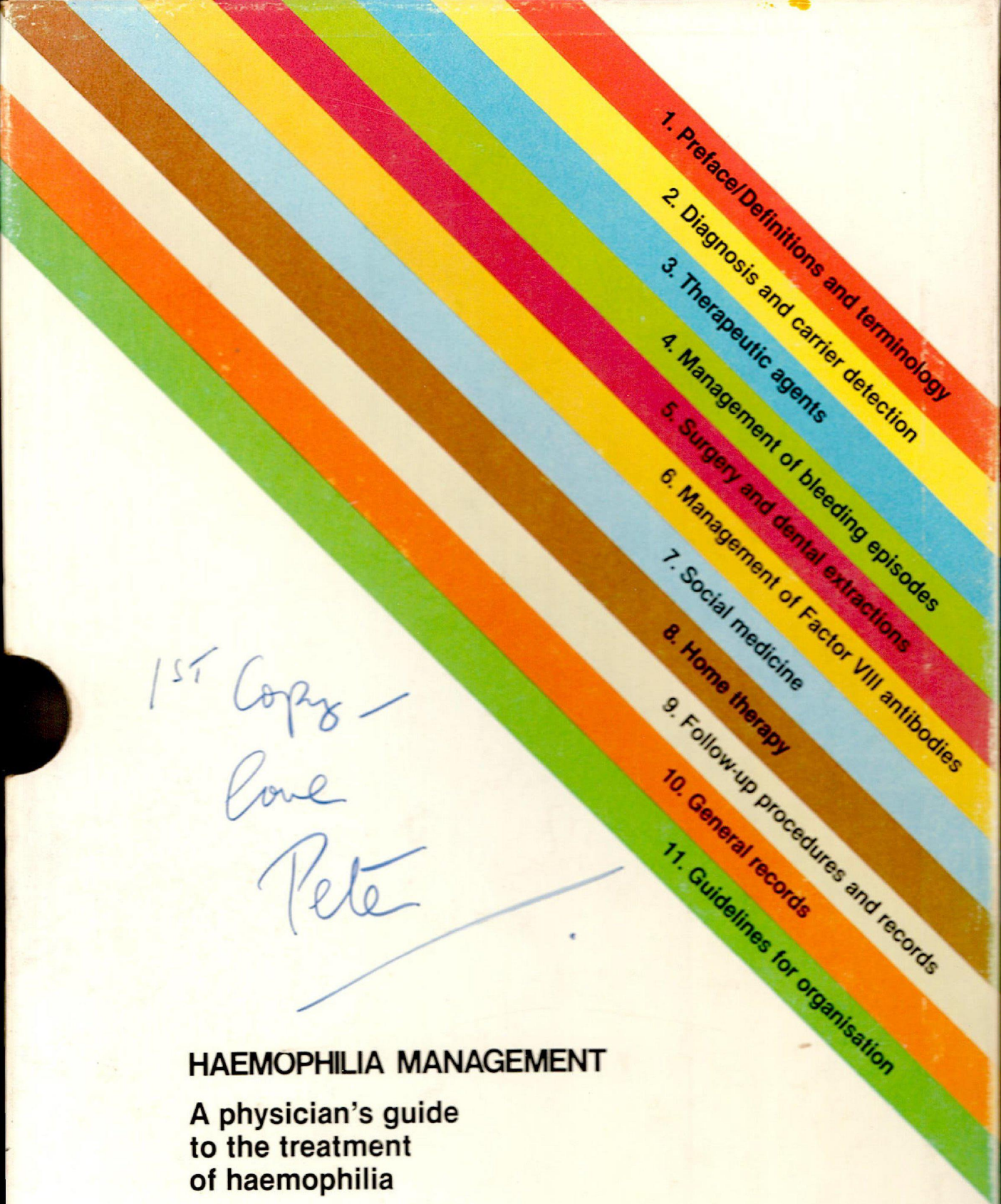


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1st Copy -
Love
Pete

HAEMOPHILIA MANAGEMENT

A physician's guide
to the treatment
of haemophilia

by Peter Jones

Designed and produced by
Transart Limited
Huntingdon
Cambs
England

1.

PREFACE/DEFINITIONS AND TERMINOLOGY

1

PREFACE / DEFINITIONS AND TERMINOLOGY

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PREFACE

The idea for this manual was developed in the course of a research attachment to the Hyland Division of Travenol Laboratories in 1977. Acutely aware of the clinician's need for concise and easily accessible information when faced with haemophilic bleeding, Hyland staff members proposed and later participated in the work. Responsibility for the medical accuracy of the contents is, however, mine, and I hope that colleagues using the manual will not hesitate to correct errors or suggest alternative methods of treatment. Their comments will be collated and distributed as supplements to the basic manual. All contributions, which will be acknowledged, should be sent to:

Judith H. Quin
Transart Limited
Huntingdon
Cambridgeshire
England.

Because the approach to haemophilia management is constantly changing in the light of new discoveries, techniques and experiences, this should be regarded as a working document rather than as a definitive text.

It is hoped, however, that readers will find it, in whole or in part, useful for teaching and as a standby for junior hospital staff and paramedical workers.

The manual is divided into eleven parts, which are colour-coded for easy reference. A list of key references is given at the end of each section, both in acknowledgement to their authors and as a guide to further reading.

I am indebted to the staff of the Newcastle Haemophilia Centre for the benefit of their expertise in the writing of parts of the work, to the staff of the Medical Illustration and Teaching Aids Laboratory of the University of Newcastle upon Tyne for the photography, and to the staff of Transart for their help in the preparation of the manual. It should be noted that copyright of photographic material is vested in the University of Newcastle upon Tyne. Parts of the work were carried out with the aid of grants from the UK Haemophilia Society and the World Federation of Hemophilia, to whom I am grateful.

Finally, without the perseverance of Robert Taub of Travenol nothing would have been achieved, and I am grateful to him for his friendship and continued advice.

Peter Jones MD FRCP DCH
Newcastle upon Tyne
1979

The study of haemostasis and coagulation is often confounded by the use of abbreviations, initials and synonyms. Many of the more common of these are included in this booklet, together with definitions of terms employed in blood product fractionation, and in laboratory and clinical work.

A

ACD	Acid citrate dextrose; anticoagulant.
Activation	The conversion of a proenzyme into an enzyme. The activated form of a coagulation factor is indicated by the subscript 'a' (Factor X _a).
ADP	Adenosine diphosphate.
AMCA	Amino-methyl cyclohexane carboxylic acid; tranexamic acid; Cyklokapron. An antifibrinolytic.
Antibody unit:	
Oxford definition	Amount which inactivates 50% of 1 unit/ml solution of factor VIII in 4 hours at 37°C.
Bethesda definition	Amount which inactivates 50% of 1 unit/ml solution of factor VIII in 2 hours at 37°C.
APPT	Activated partial thromboplastin time (≡ PTTK)
ATP	Adenosine triphosphate.
Aura	The feeling that a bleed has started <i>before</i> any outward signs are present. This early warning feeling is of the utmost importance for the success of home therapy. Bleeds treated on the recognition of an aura often stop immediately with a relatively small dose of blood product.

C

C	Coagulant activity. VIII:C = factor VIII coagulant activity.
C:Ag	The antigen related directly to factor VIII clotting activity (VIII:C). VIII:Ag may be measured either in serum or plasma by immunoradiometric assay. Unlike VIII:R:Ag it is reduced or absent in haemophilia.
CAP	Controlled activated product (Hyland). A prothrombin complex product developed for the treatment of patients with VIII antibodies.
Christmas disease	Haemophilia B. Christmas was the name of the first patient described with factor IX deficiency.
Clg	Cold insoluble globulin. Plasma protein incorporated into fibrin net by activated factor XIII.
Coagulation	The process of blood clotting, the end point of which is the appearance of insoluble fibrin.
Coagulation assay	Determination of a specific coagulation factor content of a plasma sample. In a <i>one-stage</i> assay the complete sequence of reactions to the formation of fibrin clot (the end point) takes place in a single tube, to which all the necessary reagents have been added. In a <i>two-stage</i> assay the end point (second stage) is reached in subsamples taken from a preincubation mixture of reagents (first stage).
Concentration	As applied to blood product fractionation, the ratio of activity per unit volume in the product to the activity per unit volume in the starting material.
Cryoprecipitate ('Cryo')	The cold-insoluble precipitate remaining after fresh frozen plasma has been thawed in the cold (4°C). Contains approximately 50% of the factor VIII content present in the original fresh frozen plasma, and about 30% of the original fibrinogen.

D

DDAVP	1-Deamino-8-D-Arginine Vasopressin. Synthetic derivative of vasopressin without vaso-active properties. VIII:C stimulator.
--------------	--

E

- EACA** Σ -amino-caproic acid; Epsikapron. An antifibrinolytic.
- EDTA** Ethylene diamine-tetra-acetate; anticoagulant.
- Enzyme** A specific protein catalyst. Most coagulation factors act as proteolytic enzymes in their activated form.

F

- Factor** Term given to a component of the coagulation sequence. By international convention Roman numerals designate coagulation factors, and Arabic numerals platelet factors.
- Factor VIII unit** The factor VIII activity present in 1 ml of average fresh citrated normal plasma prepared from blood collected into 3.8 per cent trisodium citrate in the proportions of nine parts of blood to one part of trisodium citrate (Biggs, 1976).
- FDP** Fibrin degradation, or digestion products; sometimes referred to as 'fibrin split products'.
- FECU** Factor eight correctional unit (Hyland Auto IX unit).
- FEIBA** Factor eight inhibitor bypassing activity (Immuno).
- FFP** Fresh frozen plasma (\equiv snap frozen plasma). Plasma removed from red cells by centrifugation and promptly frozen in an ethanol and dry ice mix.
- Fitzgerald factor** Contact factor thought to represent abnormality in high molecular weight kininogen.
- Fletcher factor** Prekallikrein.

H

- Haemophilia** Noun derived from haima (Greek = blood) and philos (Greek = loving); origin attributed to Hopff (1828), a student of Schönlein, by Brinkhous. Hence haemophilic (noun) and haemophilic (adjective). Alternative spelling hemophilia (USA), hence World Federation of Hemophilia and British Haemophilia Society.

- Haemophilia A** Synonyms: factor VIII deficiency; classical haemophilia.

A sex linked recessive bleeding disorder due to an isolated deficiency of the clotting activity of factor VIII (VIII:C).

VIII:C is the normal biological expression of a molecule containing protein with antigen sites which may be detected by immunological techniques. This protein element, factor VIII related antigen, is by convention abbreviated to VIII:Ag. VIII:Ag in haemophilia A is normal.

In addition to VIII:C and VIII:Ag the International Committee on Thrombosis and Haemostasis have prepared the following nomenclature:

VIII:WF = factor VIII related to von Willebrand factor;

VIII:WF (BT) = activities related to factor VIII associated with bleeding time;

VIII:WF (GB) = activities related to factor VIII associated with platelet retention by glass-beads;

VIII:WF (RCF) = activities related to factor VIII associated with ristocetin platelet aggregation.

Factor VIII synonyms: antihaemophilic factor (AHF); antihaemophilic globulin (AHG).

Incidence: variously estimated between one severe case per 7 000 - 12 500 males.

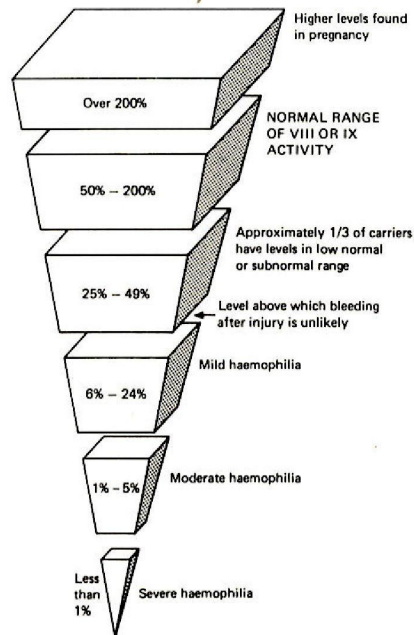
Incidence of all severities may be as great as one per 3 500 males.

Severity: normal range of factor VIII is 50 - 200%, with a mean of 100%.

Levels greater than 200% may be recorded in pregnancy.

Severe haemophilia: factor VIII < 1%.
Moderate haemophilia: factor VIII 2 - 5%.
Mild haemophilia: factor VIII > 5%.

Note: *Clinical* severity is usually in accord with VIII:C measurement but there are exceptions. In the event of major injury or surgery clinically mild cases should be treated as severe.



Inheritance: children of a haemophiliac —

Each son will be *normal*.
 Each daughter will be a *carrier*.

children of an obligatory (v.i.) carrier —

Each son has a 50:50 chance of inheriting haemophilia.
 Each daughter has a 50:50 chance of inheriting the abnormal gene and therefore being a carrier like her mother.

The severity of haemophilia in different generations of the same family tends to run true.

Obligatory carriers are —

mothers with one haemophilic son and a previous family history of the disorder;
 mothers of more than one haemophilic son;
 daughters of a haemophilic father.

Potential carriers are —

mothers with one haemophilic son and no family history;
 daughters of obligatory carriers.

Haemophilia carriers may have low VIII:C levels and be vulnerable to abnormal bleeding.

Haemophilia B Synonyms: factor IX deficiency; Christmas disease; PTC deficiency.

A sex linked recessive bleeding disorder due to an isolated deficiency of the clotting activity of factor IX (synonym: Christmas factor).

Incidence: estimated as one per 60 000 males in severe form.

Inheritance and **severity** as for haemophilia A.

Haemostasis The spontaneous arrest of bleeding from ruptured blood vessels.

Half life Term borrowed from physics to describe the time taken for the biological activity of a clotting factor to decay to half its original value.

Home therapy Treatment by the intravenous injection of the relevant blood product in the community (as opposed to within the hospital). The term covers treatment given by self-infusion or administered by another person, and treatment given at work or school or whilst on holiday, as well as at home. To be successful and safe home therapy should only be practised against a background of comprehensive care.

I

Inhibitor Term originally given to the substance which destroys a clotting factor. Many inhibitors are now known to be true IgG antibodies. Acquired specific inhibitors have been ascribed to all ten protein coagulation factors (I, II, V, VII, VIII, IX, X, XI, XII and XIII).

IRMA Immunoradiometric assay.

K

Kallikrein Enzymatic form of prekallikrein (Fletcher factor). Plasma component involved in activation of factor XII and fibrinolytic system, and in release of bradykinin.

KCCT Kaolin-cephalin clotting time (APPT).

L

Lyophilised Freeze-dried.

O

On-demand therapy Treatment given as soon as a haemophiliac thinks that bleeding has started (also referred to as crisis therapy).

P

PCI Prothrombin consumption index.

P and P method Prothrombin and proconvertin method; modification of the prothrombin time.

PEG Polyethylene glycol; fractionating agent.

Potency The terms *low*, *intermediate* and *high potency* refer to the degree of purification of a particular coagulation factor in a plasma fraction. Factor VIII is about 10 X purified (compared with fresh plasma) in a low potency product, about 16 X purified in intermediate potency products, and from 150-400 X purified in high potency products. In general terms, the more highly purified the product, the greater the likely loss of original factor and thus the poorer the yield.

PPSB Prothrombin, proconvertin, Stuart factor and antihaemophilic factor B (i.e. factors II, VII, X and IX). Term given to the blood fraction derived from EDTA plasma by Soulier et al.

Prekallikrein Fletcher factor (see Kallikrein).

Proaccelerin Factor V.

Proconvertin Factor VII.

Prophylactic therapy Prophylaxis; the regular administration of a blood product in order to maintain a level of clotting factor sufficient to prevent spontaneous bleeding. Prophylaxis may be short term or *limited* (prescribed to check recurrent haemorrhage into a particular joint or to cover physiotherapy), or prolonged \equiv *maintenance* therapy.

Prothrombin complex Collective term for factors II, VII, IX and X, all of which are vitamin K dependent.

PT Prothrombin time.

PTA Plasma thromboplastin antecedent; factor XI.

PTC Plasma thromboplastin component; factor IX.

PTT Partial thromboplastin time.

PTTK Partial thromboplastin time with kaolin (\equiv KCCT).

Purification The ratio of activity per unit weight of protein in the product to the activity per unit weight of protein in the starting material.

R

R:Ag Related antigen. VIII:R:Ag = factor VIII related antigen, assumed to be the protein content of the VIII molecule as determined by immuno-electrophoresis.

RCT Recalcified clotting time.

RIA Radioimmunoassay.

Ristocetin Antibiotic withdrawn from clinical use because of thrombocytopenic effect. Platelets from patients with classical von Willebrand's disease fail to aggregate in the presence of ristocetin; platelets from classical haemophilia A patients aggregate with ristocetin.

RVV Russell's viper venom.

S

Substrate The inactive form of coagulation factor, usually a proenzyme. Enzyme action converts the proenzyme into an enzyme (the product).

T

Target joint Haemophilic joint at risk of (further) deterioration, and at which maximum therapeutic effort should be directed.

TEG Thromboelastogram.

TGT Thromboplastin generation test.

Thrombotest Prothrombin time using a commercially prepared thromboplastin reagent.

TT Thrombin time.

V

von Willebrand's disease (VWD). A haemorrhagic disorder of autosomal inheritance characterised by both qualitative platelet and factor VIII abnormalities.

In the classical disease, inherited as an autosomal dominant, the platelet abnormality results in a prolonged bleeding time and there is a deficiency of both VIII:R:Ag and VIII:C. Recent research has established several variants of von Willebrand's disease, among them an autosomal recessive variety.

VWF von Willebrand factor.

W

WBCT Whole blood clotting time.

Y

Yield

Calculated by expressing the total amount of activity in the product as a percentage of the total amount of activity in the starting material. This gives *overall* yield. *Stage* yield is the total activity at a given stage of the process expressed as a percentage of the total activity at the previous stage.

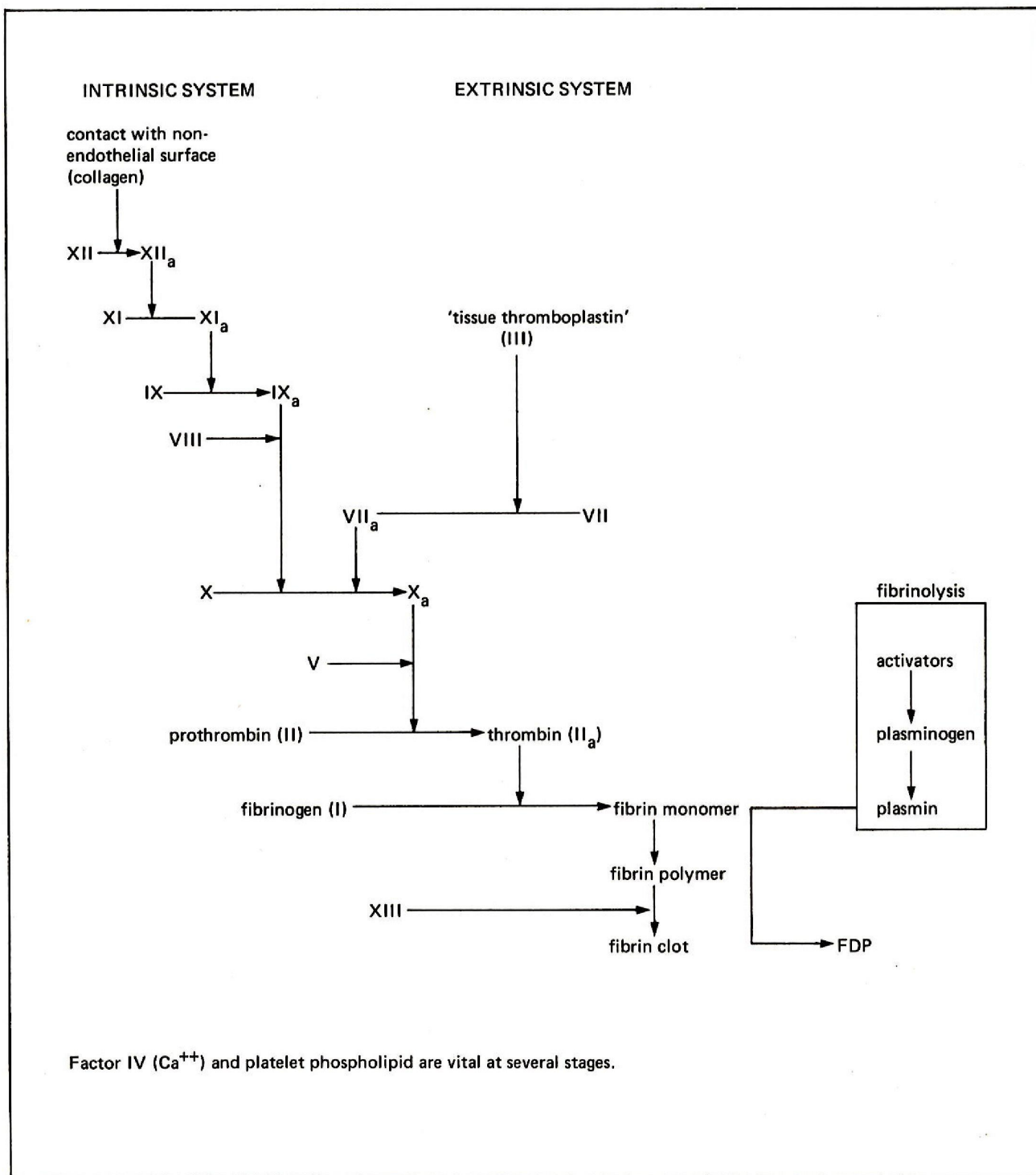
Synonyms for the Roman classification of clotting factors

- I Fibrinogen.
- II Prothrombin.
- III Thromboplastin; tissue extract.
- IV Calcium.
- V Labile factor; proaccelerin.
- VII Proconvertin; autoprothrombin I.
- VIII Antihæmophilic factor (AHF); antihæmophilic globulin (AHG); antihæmophilic factor A.
- IX Plasma thromboplastin component (PTC); Christmas factor; antihæmophilic factor B; autoprothrombin II.
- X Stuart – Prower factor; autoprothrombin III (\equiv autoprothrombin C in activated form).
- XI Plasma thromboplastin antecedent (PTA).
- XII Hageman factor.
- XIII Fibrin stabilizing factor (FSF).

Note: Christmas, Stuart, Prower and Hageman were the names of the patients from whose blood the specific defect was originally identified.

The autoprothrombin nomenclature is that of W H Seegers in *Blood Clotting Mechanisms. Three Basic Reactions*. Annual Review of Physiology 31, 269, 1969.

The coagulation and fibrinolytic pathways



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2. DIAGNOSIS AND CARRIER DETECTION

2

DIAGNOSIS AND CARRIER DETECTION

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PRESENTATION

Two thirds of haemophilia A and B patients have a family history of the disorder, and may be screened at birth.

As the factors do not cross the placenta, intra-uterine diagnosis is possible for haemophilia A, using the technique of fetoscopy; a small specimen of fetal blood is aspirated from a fetal vessel and examined for factor VIII content. The technique must still be regarded as experimental.

Now achieved for Haemophilia A – see reference

Because factor IX levels are extremely variable, the technique is not yet applicable to haemophilia B.

Fetoscopy is performed between the 18th and 20th weeks of pregnancy, in time for termination to be considered if haemophilia is diagnosed. Risk to the fetus of using this technique is currently calculated at about 5% in experienced hands.

Cord blood diagnosis may be made when an obligatory or potential haemophilia carrier is identified, provided that:

- the specimen is clean and uncontaminated with maternal blood, tissue fluid or Wharton's jelly;
- the specimen is obtained immediately on the birth of the male infant;
- the fresh specimen is tested by immediate factor assay by a competent technician.

The sample should always be inspected for clot, and the assay result should always be cross-checked with the result of the activated partial thromboplastin time (APPT; PTTK; KCCT).

Caution

Cord blood diagnosis is accurate in severe haemophilia A, but factor VIII levels may be subject to artefact in mild and moderate haemophilia, the length of gestation or mode of delivery affecting the VIII level.

Factor IX levels are very variable in the newborn and a diagnosis made on cord blood of any infant, particularly when he is not full-term and of normal delivery, must be guarded.

All infants with an initial positive diagnosis should have the relevant factor level checked in the first year of life.

Note: Diagnostic venepuncture in suspected cases of haemophilia should never be performed using either femoral or jugular veins because of the danger of haematoma formation and compression of vital structures.

Presentation of previously undiagnosed cases in early infancy is rare, usually occurring when the boy begins to crawl, pull himself up and toddle.

In rare cases presentation is with intracranial haemorrhage, usually subdural and therefore treatable, and sometimes linked with trauma.

Persistent oozing from intra-oral lesions, small cuts or anal fissures may be early presenting features.

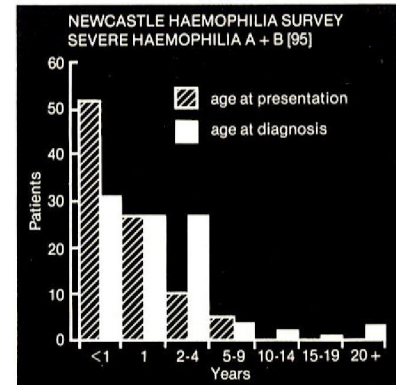
Bruising is rare before the infant is mobile — classically haemophilic bruises are raised like eggs and they spread more than bruises in normal children.

Mildly affected cases may only present after surgery — another reason for screening members of families with positive histories of bleeding diathesis.

Early diagnosis of haemophilia in the infants of identified or suspected carriers

- forewarns the family,
- prevents presentation with bleeding following circumcision or other surgery, and
- allows early counselling and treatment of bleeding episodes.

The time-lag between presentation and diagnosis which used to occur can be avoided nowadays in the majority of cases.



Ages at presentation and diagnosis of severe haemophilia A and B. From the Newcastle haemophilia survey.

DIAGNOSIS

Details of assay procedure are given in *A Laboratory Manual of Blood Coagulation* by D E G Austen and I L Rhymes (Oxford, Blackwell, 1976) and in all the standard scientific texts on haemostasis.

When confronted with a patient with a suspected haemostatic abnormality, it is useful to follow a set pattern of questioning.

HAEMOSTATIC ENQUIRY: present or past history or physical signs of

- 1 **Bruising**, purpura, telangiectasia
- 2 **Epistaxes**, GIT bleeding, haematuria, **menorrhagia**
- 3 Haemarthroses or arthropathy
- 4 Response to injury; prolonged or secondary haemorrhage after lacerations, IM injections, **dental extractions, operations, childbirth**
- 5 Blood product transfusion
- 6 Drug ingestion especially **aspirin**
- 7 **Family history**
- 8 Other disease especially **hepatic**

Suggested areas to be covered in any haemostatic enquiry.

Symptoms and signs in bold lettering are those most likely to be presenting features of a coagulopathy.

When seeing a large number of cases it is useful to adopt a formal approach to the haemostatic enquiry. Here is an example of a proforma which may be adapted for mechanical or computer sorting. The forms are reproduced in full in the booklet on *General Records*.

NORTHERN REGIONAL HAEMOPHILIA SERVICE		Diagnostic Consultation	
NAME		Telephone	
ADDRESS			
Referring Hospital/Ward/Consultant			
Study Reference		1-3	A 2
Coagulation Register Number		4-8	
Referred from: University Hospital Group = 1 G.P. = 3			
Regional Hospital = 2 Dentist = 4			
Other = 5		9	
Date of Examination		10-15	
Age (in years and months)		16-21	
Date of Birth		22	
Sex: Male = 1 Female = 2			
Employment			
G.P.			
Dentist			
Reasons for Referral:			
Bruising Yes = 1 No = 2		23	
Purpura Yes = 1 No = 2		24	
Haemorrhage Yes = 1 No = 2		25	
Swollen Joint(s) Yes = 1 No = 2		26	
Past history bleeding Yes = 1 No = 2		27	
Family history bleeding Yes = 1 No = 2		28	
Other Yes = 1 No = 2		29	
Other illness Yes = 1 No = 2		30	
Diagnosis: Established Provisional			
HISTORY			
1. Bruising: Excessive = 1 Not Excessive = 2		31	
Always associated with trauma = 1			
Sometimes spontaneous = 2		32	
Superficial bruising: Large (>5 cms. diam.) = 1			
Small (<5 cms. diam.) = 2		33	
Flat = 1 Raised = 2		34	
Deep haematoma: Experienced = 1 Never = 2		35	
Principle sites of bruising:			

CARRIER DETECTION AND GENETIC COUNSELLING

Haemophilia A and B are inherited as sex linked recessives.

Note: von Willebrand's disease is inherited in its common (classical) form as an autosomal dominant. It is occasionally autosomal recessive (therefore check for consanguinity in the family).

Obligatory carriers

Obligatory carriers of haemophilia A and B are:

- all the daughters of a haemophilic father;
- mothers with a haemophilic son and a previous family history of haemophilia;
- mothers of more than one haemophilic son.

Potential carriers

Potential carriers of haemophilia A and B are:

- mothers with no previous family history and one haemophilic son;
- daughters of known haemophilia carriers.

Carrier detection Haemophilia B (Factor IX deficiency, Christmas disease)

Carrier detection in these families is dependent on the history and the factor IX activity level, which should be measured on at least three occasions before a prognosis is given.

About 30% of haemophilia B carriers have factor IX levels below or at the bottom of the normal range (50-200%). Occasionally the level may be low enough to cause haemostatic problems on injury, surgery or dental extractions, or during menstruation or child-birth. The latter is unlikely because the level normally doubles during pregnancy.

**Haemophilia A
(Factor VIII deficiency)**

Carrier detection is theoretically possible in between 70 and 98% haemophilia A carriers, by combining the results of factor VIII biological clotting activity (or procoagulant activity) assays, FVIII:C — and factor VIII related antigen, FVIII:Ag (protein) estimations by electro-immunoassay.

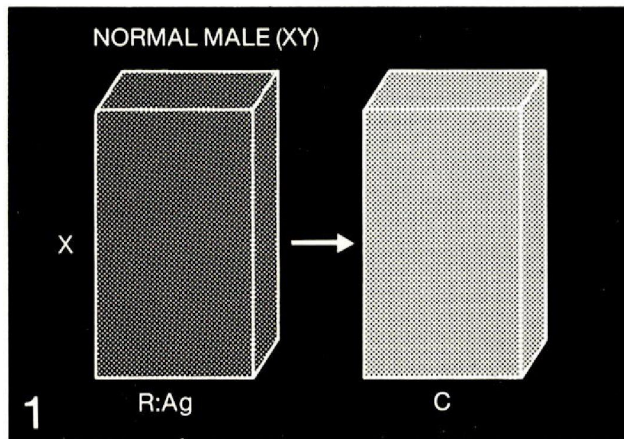
As with haemophilia B, about 30% of haemophilia A carriers have factor VIII levels in the lower normal or below the normal range. The majority also show a sufficient discrepancy between VIII:Ag and VIII:C to be identified as carriers, even when VIII:C lies within the normal range.

These diagrams show a simplified account of the rationale and of the techniques involved and have been found useful as illustrative material during genetic counselling of families.

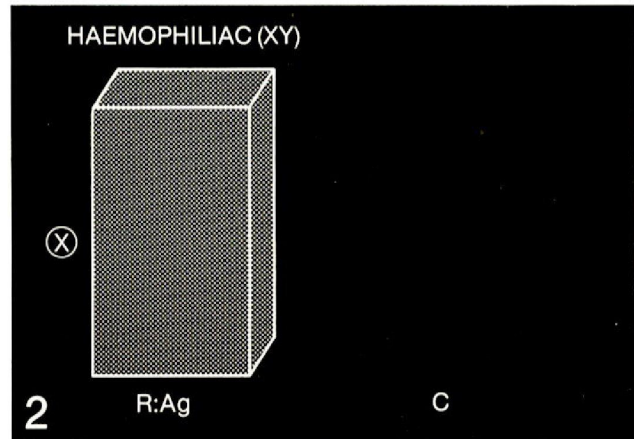
The techniques have been the subject of a study by the World Health Organization and it is currently recommended that

at least three specimens are taken from the non-pregnant subject, with at least two weeks between sampling.

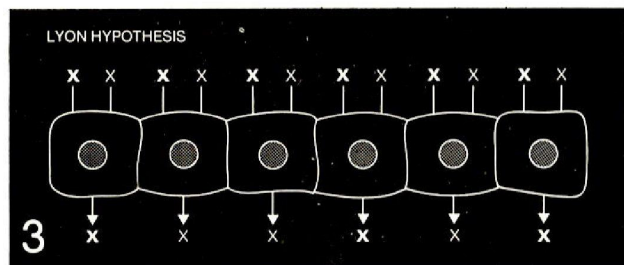
The specimens should preferably be taken at the same time of the day in the resting state and the subject should be free from illness at the time of sampling.



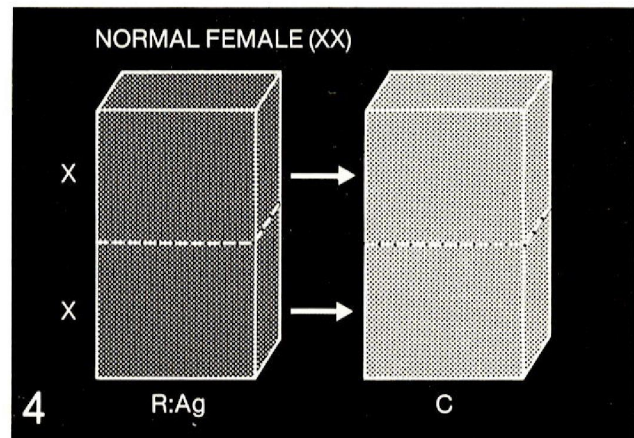
In the normal male the gene on the factor VIII locus of the X chromosome produces an immunologically detectable factor VIII related antigen (VIII:R:Ag) with equivalent normal biological activity (VIII:C).



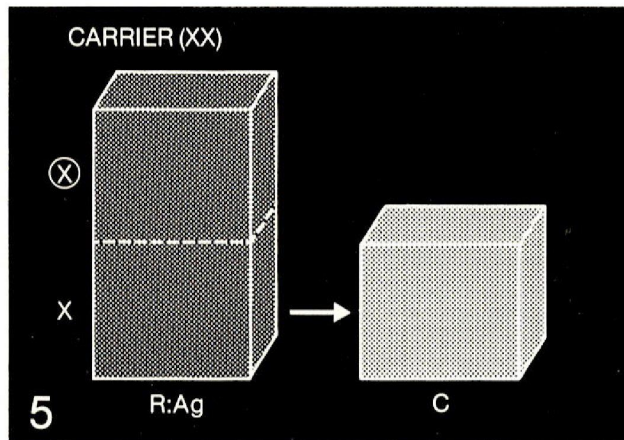
In the case of the haemophiliac, an immunologically detectable protein (controlled by the defective X-linked gene) produces no biological activity.



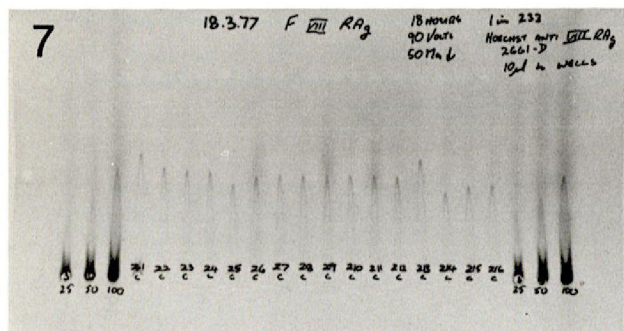
The Lyon Hypothesis: of the two X chromosomes present in each body cell of the female only one is active (the other becoming the Barr body). Thus in the average case 50% of the factor VIII (or IX) production will be controlled by one X chromosome (from one parent) and 50% by the other X chromosome (from the other parent). In cases of extreme Lyonisation one X chromosome is predominant and if this carries a defective factor VIII gene the factor VIII activity level (VIII:C) of the female will be abnormally low.



In the average normal woman one set of X chromosomes produces 50% of the factor VIII related antigen and thus 50% of the factor VIII activity. The other set of X chromosomes (derived from the other parent) produces the other 50% VIII:R:Ag and VIII:C.



The haemophilia carrier: upper X chromosome in diagram bears haemophilia gene — immunologically detectable protein but no activity result; other X chromosome bears normal factor VIII genetic locus with resultant normal activity.



Laurell Immuno-electrophoresis for factor VIII related antigen (VIII:Ag; VIII protein). The three wells at either end of the gel contain dilutions of a standard. The remaining wells contain test plasmas.

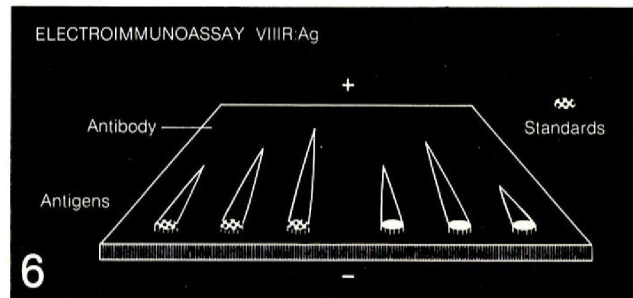
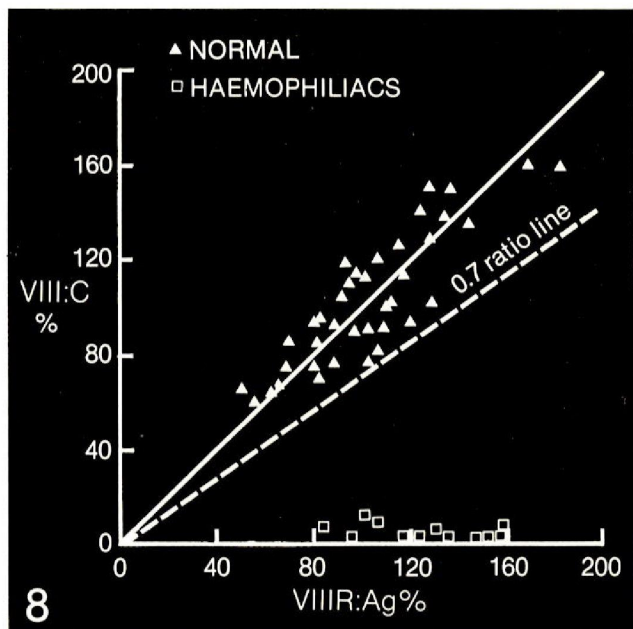
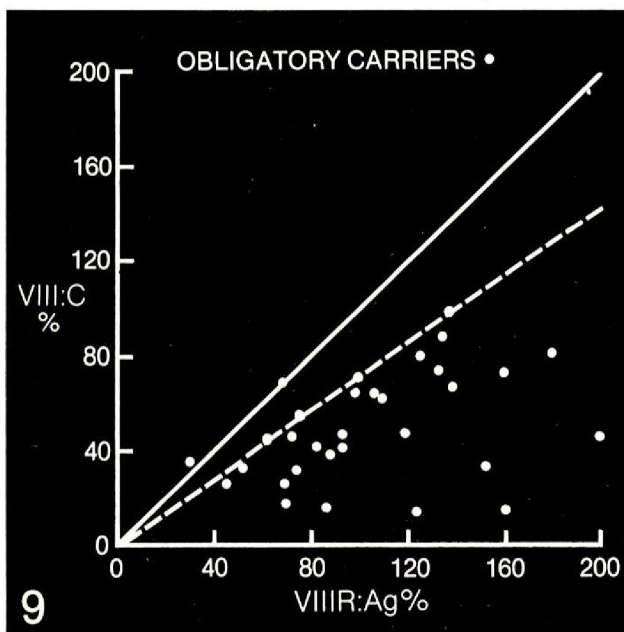


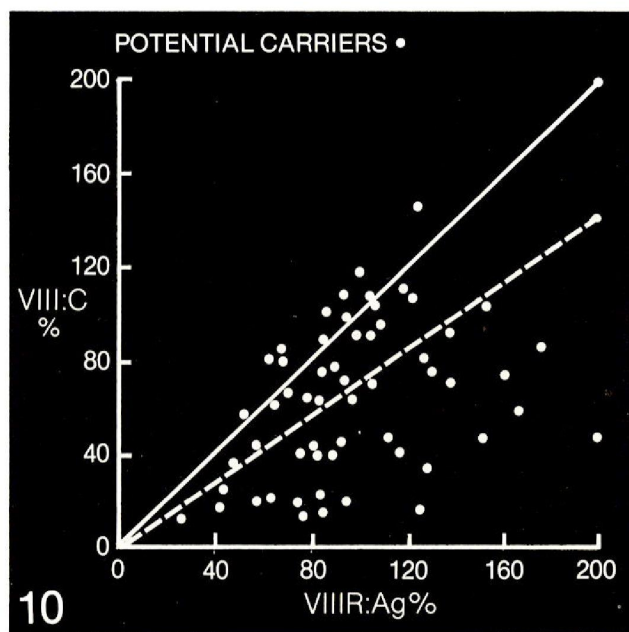
Diagram of electroimmunoassay (Laurell). The gel contains the factor VIII antibody (raised in a rabbit by injecting human factor VIII) and the wells contain the factor VIII antigen. The three wells on the left contain different dilutions of a standard — from the results obtained on these samples, the amounts of factor VIII related antigen in the other samples can be determined. When a current is passed through the gel the contents of the wells are drawn out, producing rockets of antigen which react with the antibody. The height of each rocket is directly proportional to the amount of antigen present in the sample.



Haemophilia A carrier detection.



Haemophilia A carrier detection. 0.7 ratio VIII:C to VIII:Ag used as cut-off point in Newcastle Centre. Majority of obligatory carriers fall on or below this line, *but not all*. Note in particular the two obligatory carriers on the normal (50:50) line. This is why it is not possible to tell a girl that she is definitely *not* a haemophilia carrier.



Haemophilia A carrier detection. Graph shows results on a series of *potential* haemophilia carriers. As expected from the Lyon Hypothesis, about half fall within the normal range for this particular laboratory.

Detailed discussion of the methods presently available for haemophilia carrier detection is available in the *Joint Project Report of the World Health Organization and the World Federation of Hemophilia*, which is to be found in the *Bulletin of the World Health Organization* 55(6): 675-702 (1977), and from which the following extract is reproduced.

PRIMARY RECOMMENDATIONS

1. The organization of facilities for carrier detection should be included in each country's national haemophilia programme. Carrier detection should be made available to those who require it.

2. All laboratories entering into carrier detection should be thoroughly experienced in the technology of the assays, which will be based on the local or working standard calibrated ultimately with the International Standard. Careful pedigree analysis and recognition of carriers with a deficiency of factor VIII or IX will decrease the number needing detection analysis.

3. Every country should aim to establish its own national standard for factor VIII, calibrated in terms of the International Standard. In some cases it may be more appropriate for groups of countries to combine in providing national standards. At present an International Standard is available only for factor VIII coagulant activity (factor VIIIIR:C). It is recommended that a standard should be made available for factor VIIIIR:Ag.

4. Methods for the detection of haemophilia carriers demand that laboratories be developed in regions that have a large haemophilia population and/or the technical expertise to ensure accuracy. When the technology is mastered it is important to study at least 30 obligatory carriers, preferably unrelated, to ensure the ability to detect them as compared to a similarly sized reference group of normal women. It is imperative that statistical help in the form of a biostatistician and/or geneticist be obtained.

5. A country should ensure that personnel are trained appropriately and then given the opportunity to have their data evaluated. This may well be accomplished via the ongoing workshop programmes of the International Hemophilia Training Center Committee of the World Federation of Hemophilia.

6. When the data are available it is imperative that they be presented in a way that the patients can understand. This may be done by a variety of people — the specialist physician with experience in the care of haemophiliacs, the primary care physician, or the trained genetic counsellor. The psychosocial impact of the information must be given major consideration and the counsellor, either in isolation or as a member of a comprehensive care team, needs to be prepared to give support to the patient and family.

7. Little is known about the success of the education process and we encourage evaluation of this in all those counselled.

8. A careful follow-up must be made of the outcome of pregnancies in possible carriers who have been assessed in the carrier detection programme. The offspring will either be normal or haemophilic males, or carrier females. The accuracy of each carrier detection centre can be monitored by comparing, for each woman, the predicted risk for being a carrier with the clinical status of the children who are subsequently born.

SECONDARY RECOMMENDATIONS

There remain several unresolved but important issues. We encourage investigators to attempt to solve some of these.

1. The effect of pregnancy and oral estrogen intake on carrier detection is unknown. It is important to determine whether separate carrier and control groups are needed for such patients.

2. At present almost all carrier data are being determined for members of pedigrees where severe haemophilia has been defined. It is of interest and potential importance to establish reference data for obligatory carriers of moderate and mild haemophilia also.

3. Further research is required on the antenatal diagnosis of haemophilia in males.

4. More precise information is needed on the prevalence of haemophilia in the population, together with the fitness and mortality rates for affected individuals. Only in this manner can the future prevalence of haemophilia and haemophilia carriers be accurately projected.

5. Development of better laboratory methods for the detection of haemophilia B carriers is required.

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3.

THERAPEUTIC AGENTS

3

THERAPEUTIC AGENTS

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BLOOD PRODUCTS

Fresh frozen plasma Although few, if any Centres in developed countries now use fresh frozen plasma in the treatment of haemophilia A and B, it is useful in certain clinical situations and is the source material for other products.

Preparation This product is prepared either by separating the plasma from the cells within a specified time (usually between 6 and 18 hours) of donation, or by plasmapheresis.

Storage FFP should be stored in a deep freeze at -30°C (maximum).

Shelf life In these conditions it will last at least six months. Once FFP has been thawed out it must be used immediately.

Preparation for use The product is thawed by immersing the whole plastic pack in water at 37°C . The pack is then wiped dry and examined for defects such as leaks, before the plasma can be considered safe for use.

Administration Administration is by intravenous drip.

ABO and Rhesus blood groups should be compatible for the patient, as cellular (antigen) material may be present. This is especially important when transfusing Rhesus negative females of prepubertal or childbearing age.

The rate of transfusion depends on the blood volume, but FFP should be given as fast as possible; long transfusion time lowers *in vivo* clotting factor peaks.

Side effects Allergic reactions such as chills, rigors and urticaria are common, especially in multi-transfused patients.

It is recommended that the antihistamine, chlorpheniramine (5 mg children; 10 mg adults), be given intravenously at the start of transfusion.

Acute allergic pulmonary oedema may occur during or within 24 hours of therapy with FFP or cryoprecipitate. This is believed to be due to white cell antibodies or anti-Gm (1) precipitins in the donor plasma.

Patients may complain of praecordial tightness, acute chest pain (mimicking myocardial infarct pain), epigastric pain, dyspnoea or backache. Pyrexia and cyanosis may be present, but not invariably.

An expiratory wheeze and/or fine crepitations may be heard on auscultation. Occasionally there is tachycardia.

Diagnosis is on chest X-ray appearance of pulmonary oedema with multiple scattered opacities, in the absence of evidence of circulatory overload.

Treatment is with immediate intravenous hydrocortisone (100 mg) and frusemide (40mg), repeated 4–6-hourly for 24 hours.

It seems sensible to treat patients who have once developed acute allergic pulmonary oedema with concentrates rather than with FFP or cryoprecipitate in the future.

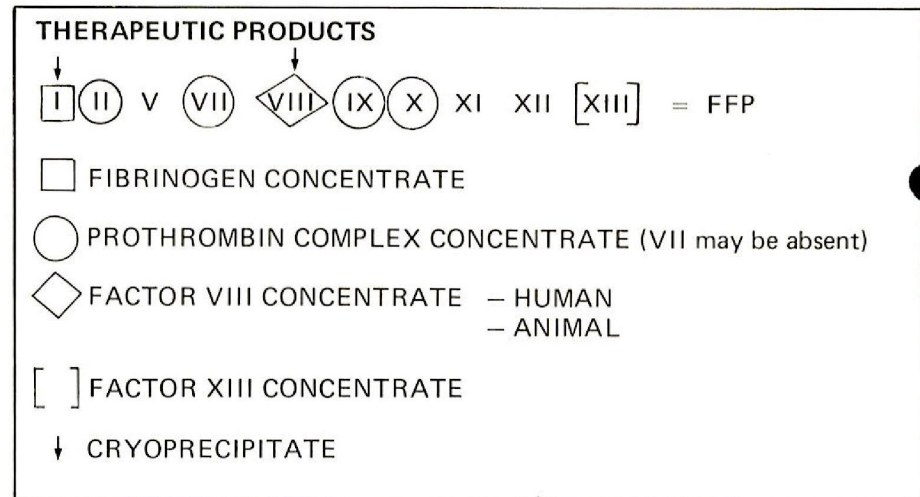
Because fresh frozen plasma is in single donor packs, the risk of transmitting serum hepatitis is small, *but it is still present*.

Use FFP contains all known protein clotting factors and in all probability other, as yet unidentified, active coagulation agents.

It is, however, a product of such low potency that it is *only* of use for the treatment of *minor* bleeds in haemophilia A or B, since levels of greater than 20% are impossible.

Attempts to achieve higher *in vivo* levels may result in hypervolaemia and congestive heart failure.

FFP is at present the only product for the treatment of bleeding in people with deficiencies of factors V and XI. It may also be used in deficiencies of factors II, VII, X and XIII, although concentrates of these factors are more convenient.



FFP is occasionally prescribed for those cases in which abnormal haemostasis cannot be explained by faulty surgical technique, infection, or abnormal laboratory results.

Cryoprecipitate Preparation

This product is prepared from fresh frozen plasma thawed in the cold at 4°C. The resultant precipitate is separated from most of the residual plasma in a leaf press after centrifugation.

The residual plasma is a source material for other proteins, including albumin, gamma globulin and the prothrombin complex.

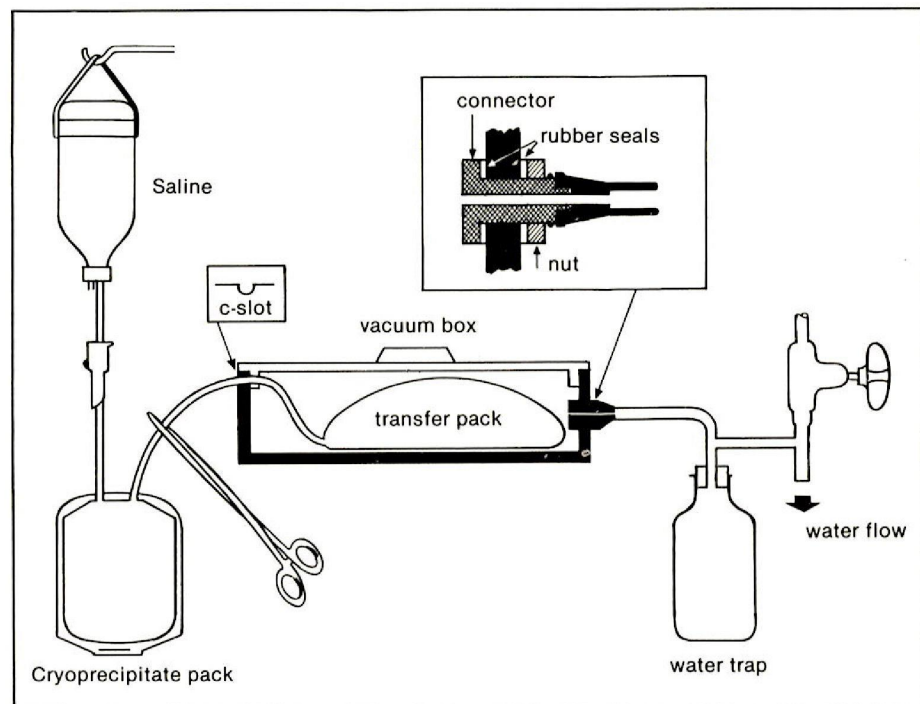
Storage It is stored in a deep freeze, preferably at -30°C (maximum).

Shelf life Under these conditions cryoprecipitate will last for at least 6 months. Once it is thawed it must be used immediately.

Preparation for use As cryoprecipitate is prepared and stored in individual donor packs, it must be pooled before use (unless a small-dose injection is to be administered to an infant, when individual packs may be thawed and given by syringe and filter).

The packs are thawed at 37°C and the contents pooled. Each pack should be washed out with a small volume of sterile isotonic saline to ensure the removal of the maximum volume of factor VIII.

Diagram of a simple and inexpensive device for pooling cryoprecipitate. The vacuum box is evacuated by means of a Venturi pump worked from an ordinary water tap.



Administration Administration is by intravenous filtered drip. Cryoprecipitate should be Rhesus compatible when administered to women of reproductive or pre-reproductive age. A small volume of pooled cryoprecipitate and washings allows a fast transfusion rate, even in children.

Side effects As with FFP, immediate allergic reactions are possible and it is recommended that antihistamine be given as prophylactic routine.

Also as with FFP, *acute allergic pulmonary oedema* is a possible side effect.

There is the long-term risk of serum hepatitis.

Use This product is for use in haemophilia A (factor VIII deficiency) and in von Willebrand's disease.

Cryoprecipitate does *not* contain factor IX.

Although it may be used as a source of fibrinogen, cryoprecipitate should be conserved for factor VIII therapy, as far as is practicable.

CRYOPRECIPITATE: APPROXIMATE VIII:C UNITS IN n PACKS

Number of packs needed	Mean VIII:C Yield (Units) per pack				
	50	60	70	80	90
2	100	120	140	160	180
3	150	180	210	240	270
4	200	240	280	320	360
5	250	300	350	400	450
6	300	360	420	480	540
7	350	420	490	560	630
8	400	480	560	640	720
9	450	540	630	720	810
10	500	600	700	800	900
11	550	660	770	880	990
12	600	720	840	960	1080
13	650	780	910	1040	1170
14	700	840	980	1120	1260
15	750	900	1050	1200	1350
16	800	960	1120	1280	1440
17	850	1020	1190	1360	1530
18	900	1080	1260	1440	1620
19	950	1140	1330	1520	1710
20	1000	1200	1400	1600	1800

The figures in the box show the total number of factor VIII units expected according to the mean of the measured VIII:C yield.

Note: Allowances should be made for the increased chance of lower dosages being diluted by occasional packs of poor yield.

Minimum dose in infants should be 2 packs.

Actual dosage depends on patient's response. This should be measured by factor VIII assay when repeated doses are required for management of severe haemorrhage or surgery.

The factor VIII:C content of the pool is increased by rinsing out cryoprecipitate packs with 0.9% sterile saline.

**Lyophilised concentrates
Preparation**

Fresh frozen plasma is treated with organic solvents or amino acids to produce intermediate purity products. Further treatment with polyethylene glycol (PEG) produces higher purity concentrates.

Factor II, VII, IX and X concentrates may be prepared from plasma or cryoprecipitate supernatant.

AHF CONCENTRATE FLOW CHART

CRYOPRECIPITATE

PLASMA



CRYOPRECIPITATE
FORMATION



CENTRIFUGE



CRYOPRECIPITATE

INTERMEDIATE POTENCY

PLASMA



CRYOPRECIPITATE
FORMATION



CENTRIFUGE



CRYOPRECIPITATE
PURIFICATION



LYOPHILISATION



INTERMEDIATE POTENCY
CONCENTRATES

METHOD FOUR (HYLAND)

PLASMA



CRYOPRECIPITATE
FORMATION



CENTRIFUGE



CRYOPRECIPITATE
PURIFICATION



POLYETHYLENE GLYCOL
(PEG) PURIFICATION
AND CONCENTRATION
PROCEDURE



GLYCINE PURIFICATION
PROCEDURE



LYOPHILISATION



HEMOFIL
CONCENTRATE

Storage

This varies according to the product but it is usually at $4^{\circ}\text{C} \pm 2^{\circ}\text{C}$, or at room temperature for specified periods of time up to 6 months.

Shelf life

The shelf life is usually 1-2 years, depending on the product.

Preparation for use	<p>A diluent (distilled sterile water for injection) is added to the lyophilised concentrate, using an aseptic technique.</p> <p>In order to ensure the removal of potentially harmful particulate matter, a filter needle should always be used to draw up the solution for injection.</p> <p>The exact method of preparation depends on the product, as does the time it takes for the concentrate to go into solution — this varies from a few seconds to 45 minutes.</p> <p>Usually warming to 37°C (both of the concentrate bottle and the diluent), introduction of the diluent slowly to avoid lumps, and gentle rotation avoiding frothing, are advised.</p> <p>Reconstituted concentrates should be used within an hour of mixing.</p>
Side effects	<p>Immediate side effects are extremely rare, especially with the high purity products.</p> <p>Intermediate purity products occasionally produce <i>allergic</i> type reactions.</p> <p>High dosage occasionally results in <i>haemolytic</i> reactions, due to blood group iso-haemagglutinins.</p> <p>Prothrombin complex products may be <i>thrombogenic</i>, and should not be used in the treatment of neonates.</p> <p>Although most commercial plasmapheresis donations are now screened for Hb_sAg by RIA or other sensitive methods, all concentrates prepared from large donor pools carry a greater risk of serum hepatitis and possibly other disease transmission than cryoprecipitate. They should therefore be reserved for the treatment of severe haemophilia A in older children and adults. Cryoprecipitate is the material of choice for young children and patients with mild haemophilia A.</p> <p>Long-term side effects involving altered immunity and renal and hepatic dysfunction have been postulated in the multitransfused population.</p>
Use	<p>Lyophilised concentrates are ideal for <i>home therapy</i> in both haemophilia A and B patients, and for surgery and high dose antibody treatment.</p> <p>They have the advantage of a known dose and a small volume for injection by syringe.</p> <p>They are, however, possibly of less value in the treatment of von Willebrand's disease, as they do not contain the bleeding time factor.</p>

Note: A summary of *Coagulation Factor Concentrates* produced by major manufacturers in Europe, America and Australia, prepared in 1978, may be found in:
 Smith J K and Bidwell E.
Therapeutic materials in the treatment of coagulation defects
 Clinics in Haematology 8, 183, 1979.

CRYOPRECIPITATE AND CONCENTRATES: ESTIMATED DOSAGE IN VIII:C UNITS

WEIGHT OF PATIENT IN Kg	RISE VIII:C WANTED %					
	15	30	45	60	75	90
10	100	200	300	400	500	600
20	200	400	600	800	1000	1200
30	300	600	900	1200	1500	1800
40	400	800	1200	1600	2000	2400
50	500	1000	1500	2000	2500	3000
60	600	1200	1800	2400	3000	3600
70	700	1400	2100	2800	3500	4200
80	800	1600	2400	3200	4000	4800
90	900	1800	2700	3600	4500	5400
100	1000	2000	3000	4000	5000	6000

The figures in the box show the number of factor VIII units required.

Formula for calculation

$$\frac{\text{WEIGHT (Kg)} \times \text{RISE VIII WANTED (\%)}}{\text{UNITS IN DOSE}} = K$$

K represents the rise in VIII:C in the patient's blood for every unit of transfused VIII/Kg body weight.

K: for plasma = 2

K: for cryo/concentrate = 1.5

Note: In our experience K may be rather lower for the initial dose (perhaps reflecting increased uptake at a site of haemorrhage or tissue dispersal).

Actual dosage depends on patient response and on timing of the dose. Bleeds caught early usually respond with lower doses than established bleeds.

In haemophilia B, K varies between 0.7 (citratd plasma or derived product) and 1.2 (PPSB type products).

Factor VIII animal products (porcine or bovine AHG)

These are nowadays reserved for the treatment of patients with factor VIII antibodies.

They are themselves antigenic, having a useful therapeutic span of only about ten days.

See Rizza C R. *The management of patients with coagulation factor deficiencies*. In *Human Blood Coagulation, Haemostasis and Thrombosis*. Ed. Biggs R, Chapter 13, 374. London, Blackwell 1976.

They also induce thrombocytopenia by platelet aggregation. Platelet counts should therefore be performed as routine during their use.

Rigors are more commonly encountered than with the human concentrates.

Note: These products are unavailable in some countries.

OTHER THERAPEUTIC AGENTS

DDAVP

1-Deamino-8-D-Arginine Vasopressin has been used to raise the factor VIII level in patients with moderate and mild haemophilia, and in those with von Willebrand's disease. Its use can thus help to conserve blood products for more severely affected haemophiliacs.

It has no value in severe haemophilia; the recipient must be capable of producing some normally active factor VIII *in vivo*.

Dosage 0.4 mg/Kg body weight, either daily or 12-hourly.

Restrict fluid intake to essentials (the patient should only drink when thirsty) and monitor for water overload and hyponatraemia.

Antifibrinolytics

Epsikapron (EACA: Epsilon amino caproic acid) and *Cyklokapron* (AMCA: tranexamic acid) may both be given orally or intravenously. They act by preventing the breakdown of a formed clot.

Use They are of value in overt haemorrhage in haemophilia A and B and von Willebrand's disease, especially in the management of dental extraction, and of epistaxis, when they may be administered by the intranasal application of the solution intended for intravenous use by nasal dropper or spray.

Dosage EACA 0.1 g/Kg body weight 6-hourly.
(intravenous or oral) AMCA 0.5g to 1.5g 8-hourly.

Side effects These include gastrointestinal upset (more common with EACA) and transient postural hypotension.

Mental confusion has been attributed to EACA in the elderly.

Contra-indication Antifibrinolytics are *contra-indicated in the presence of haematuria* or in cases of renal impairment. They predispose to clot formation in the renal pelvis, with consequent colic as the clot is passed, and the danger of renal tract obstruction.

Local haemostats These are usually of little value as they are quickly washed away by the bleeding.

An antifibrinolytic applied intra-nasally may, however, control epistaxes (v.s.), and topical thrombin may be useful in small open wounds or in dental extractions, when it may be applied on a pledget of absorbable material such as *Sterispon* or *Oxycell*.

A paste of factor VIII or IX concentrate applied locally has proved useful in patients with antibodies.

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4. MANAGEMENT OF BLEEDING EPISODES

4

MANAGEMENT OF BLEEDING EPISODES

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THE BASIC RULES OF TREATMENT

Treat bleeds early

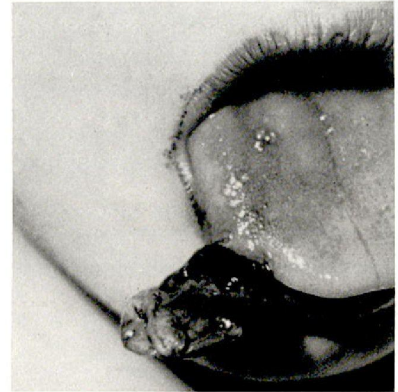
Do not wait for the appearance of physical signs.

Note: From the age of about six years haemophilic children can recognise internal haemorrhage very early in its course. This premonitory feeling or 'aura' is very valuable; treatment given at this stage will often terminate the bleed before tissue damage occurs. Less therapeutic agent is needed and activities are not interrupted.

If in doubt, treat

If a haemophiliac has sustained an injury or if he thinks he may be bleeding, treat.

The first dose of therapeutic material should be too large rather than too small.



Result of inadequate replacement therapy. Friable clot and granulation tissue extend from a tongue bite in a boy with severe haemophilia A.

Care for the veins

Never cut-down, except in a dire emergency. A haemophiliac's veins are his life-line; cut-down destroys them and poor venepuncture technique damages them.

Ban intramuscular injections (see colour plate 1)

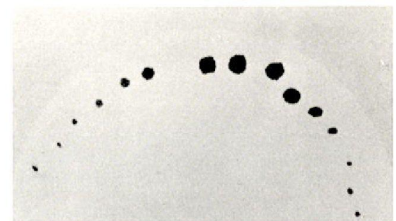
Haemorrhage provoked by intramuscular injections may result in necrosis, fibrosis, compression of vital structures and crippling.

Note: Immunisation is safe, provided that pressure is applied to the site of injection for at least five minutes. If an intramuscular bleed is provoked, it should be treated in the usual way. Immunisation apart, it is safer to ban all intramuscular injections, including pre-medication, and not to rely on blood product cover.

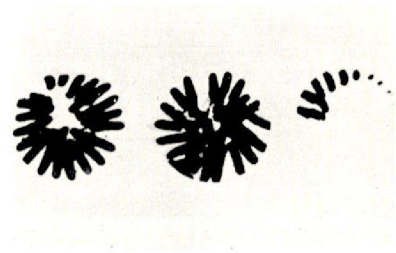
Ban aspirin

Aspirin (ASA: acetylsalicylic acid) adversely affects platelet function and induces gastrointestinal haemorrhage.

Note: All drugs carrying a risk of peptic ulceration or platelet dysfunction should be avoided. In addition to the salicylates the list includes phenylbutazone, oxyphenbutazone and indomethacin.

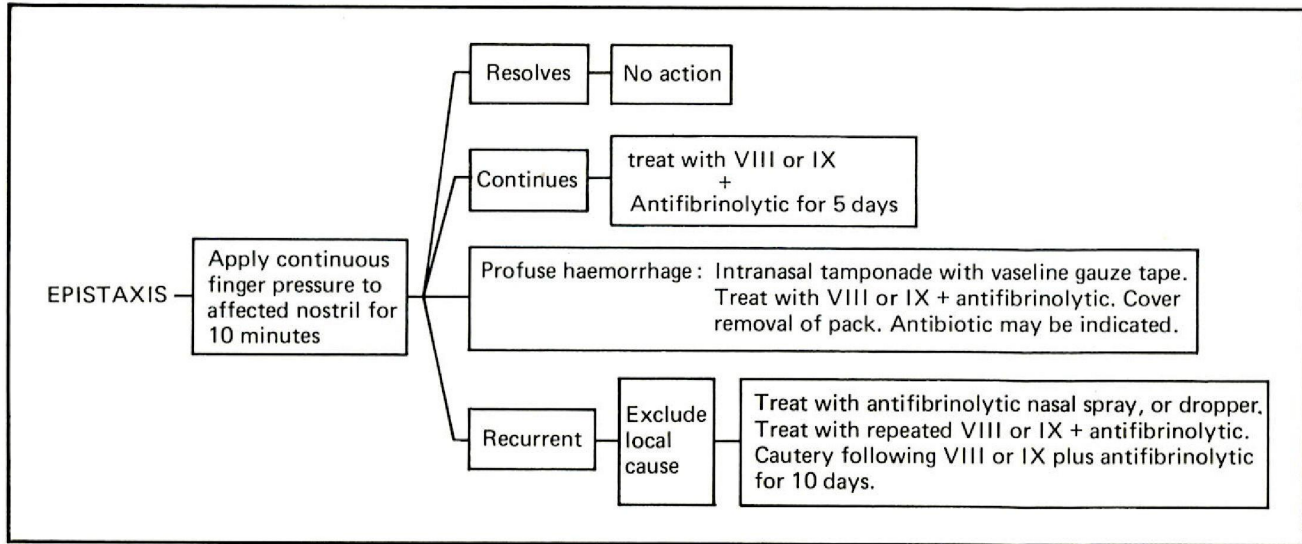


Duke bleeding time before and after aspirin ingestion. Patient was a boy who had haemorrhaged after dental extractions, having been given aspirin for pain prior to the procedure.



OPEN BLEEDS

Epistaxis

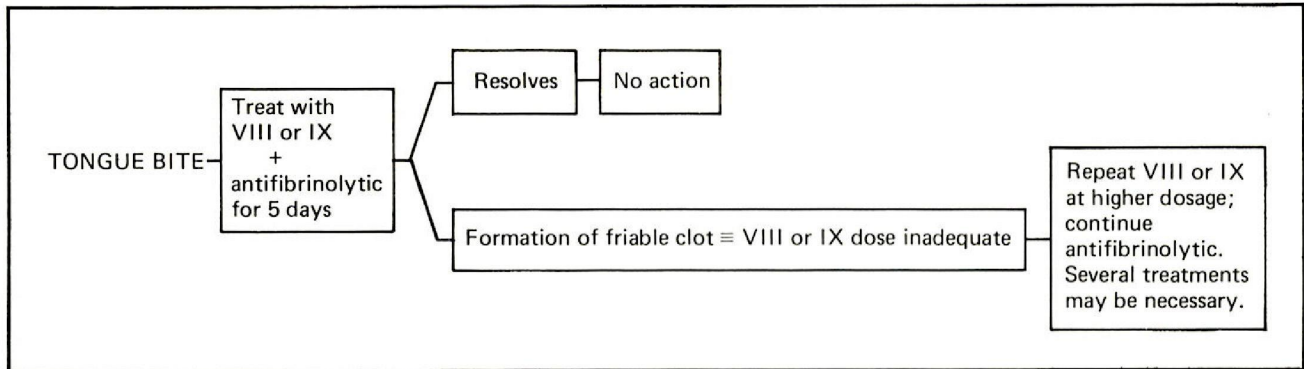


If bleeding is always from one nostril, suspect a local cause. The commonest causes of epistaxes are inflammation associated with colds, and nose picking.

Both cautery and methods of intranasal tamponade carry the disadvantage of possible secondary haemorrhage. Cautery is indicated for localised lesions — usually in Little's area — which bleed frequently and recurrently; antifibrinolytic therapy should be provided until healing is complete. Tamponade, only indicated for profuse haemorrhage, should be performed under blood product cover. A factor VIII or IX level of over 25 per cent should be attained before the pack is removed.

Intranasal spray, using the intravenous preparation of tranexanic acid, is sometimes useful in intractable recurrent epistaxes.

Oral bleeding



Persistent oozing from a small tear of the frenum may be the presenting feature of haemophilia in small children; one dose of blood product together with an anti-fibrinolytic syrup for five days is usually effective therapy. The same treatment should be used for lesions of the hard palate and buccal membranes. Lesions of the soft palate should be treated with repeated VIII/IX and an antifibrinolytic because of the dangers of respiratory obstruction. Tongue bites treated early usually resolve with one treatment, followed by an antifibrinolytic.

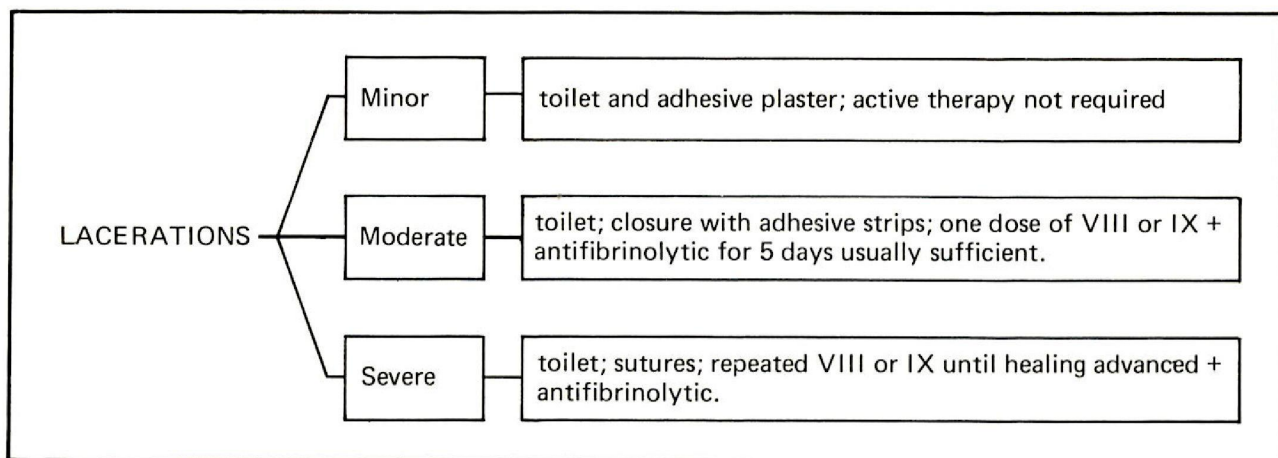
Bleeding during exfoliation of deciduous teeth is rarely persistent or dangerous. Very loose teeth should be removed; usually no further treatment is necessary.

Harsh foods such as nuts, toast, crisps, chips and biscuits should be avoided until healing of an oral lesion is complete. This takes three to five days.

Facial wounds

In children and females with a haemostatic defect facial wounds should be treated energetically because of the risk of scarring. Treat twelve-hourly with VIII/IX for at least three days or, in major lesions, until healing is complete.

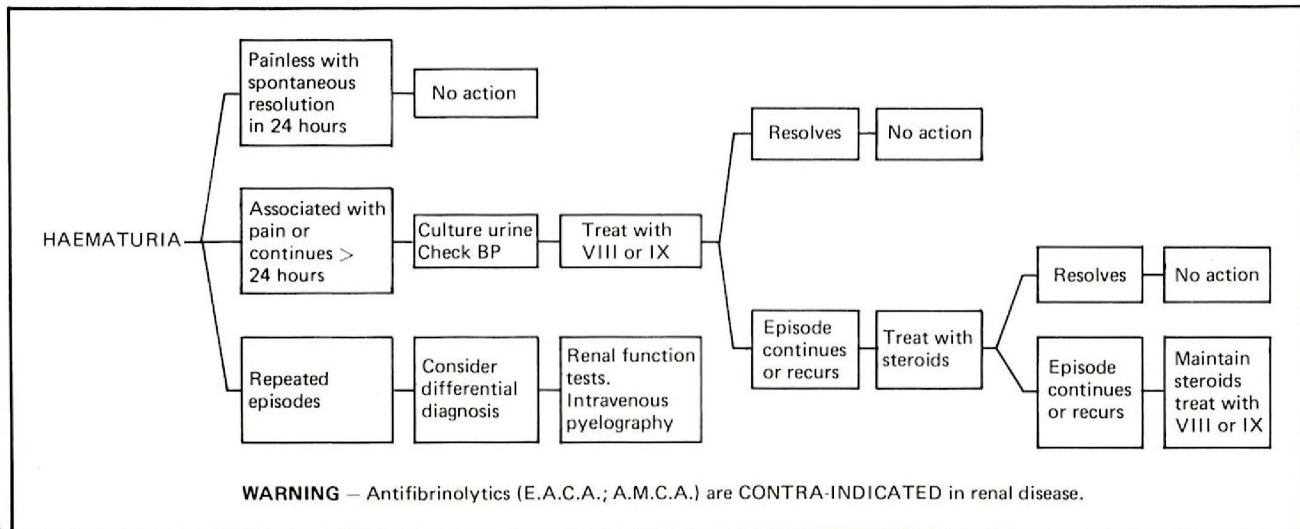
Lacerations



Check tetanus prophylaxis; antibiotic may be indicated. Close the wounds in the routine way but do not draw the edges together too firmly; tissue ischaemia may result in secondary haemorrhage. If a dressing other than adhesive plaster is indicated, use a non-stick material as the first layer. A pressure bandage should be applied after closure of major or bruised wounds.

Haematoma formation at the site of an open wound should as a rule be treated conservatively with VIII/IX cover; attempts at exploration or evacuation, except when a foreign body is suspected, are contra-indicated.

Haematuria



Antifibrinolytic strictly contra-indicated. If bleeding is from the kidney, clots will form in the renal pelvis with consequent obstruction and renal colic.

Exclude infection. Encourage fluid intake. Check BP. If haematuria is persistent or recurrent, consider differential diagnosis. Intravenous pyelography is indicated in all cases after three episodes of haematuria.

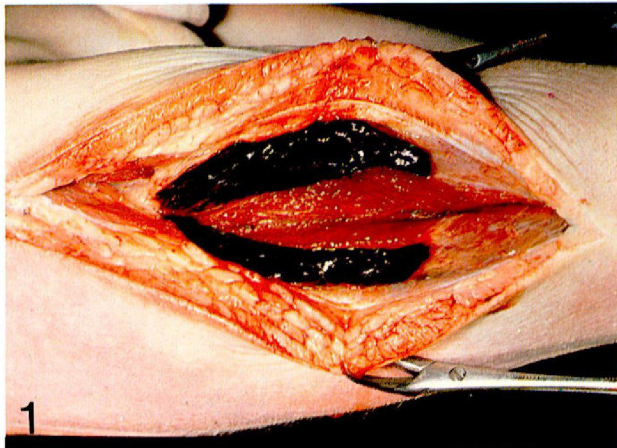
Steroid dosage for haematuria: initial dose for adults, 60 mgs (children, 40 mgs), decreasing to nil over five days.

Haemospermia is an occasional complaint in haemophilia; the commonest cause is hypertension.

Haematemesis/melaena

Investigation and general treatment are as per medical and surgical routine. Check for prior alcohol or aspirin ingestion.

Except in cases of occult blood loss under investigation, treatment with VIII/IX is indicated and should be continued until the danger of acute haemorrhage has passed. Oral antifibrinolytics are also indicated. *Fresh* blood replacement is unnecessary, provided that VIII/IX is being given; ordinary bank blood or packed red cells are more easily obtainable. It is wise to perform an antibody screen prior to clotting factor therapy, in case surgery is needed, and to check the platelet count.



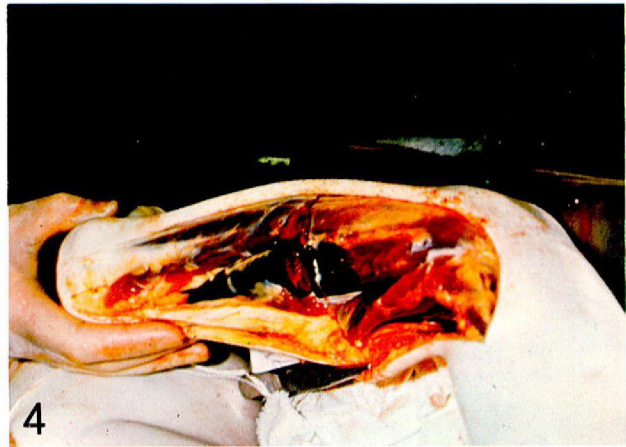
1
Extensive haemorrhage resulting from intramuscular injection into lateral aspect of thigh. Autopsy photograph.



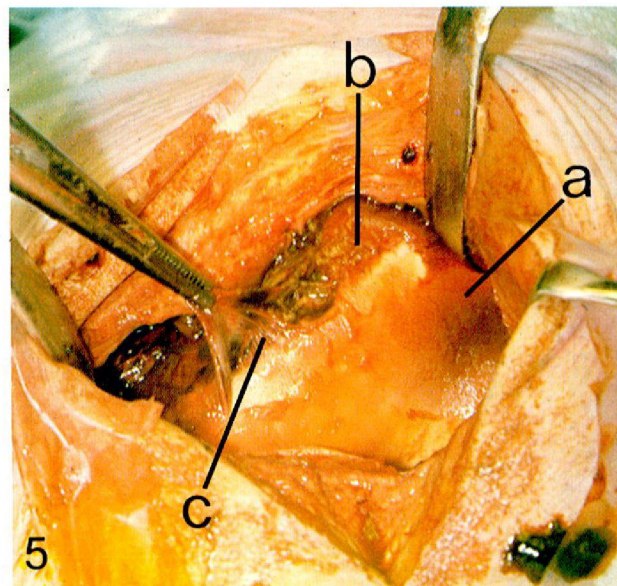
2
Spreading bruise with raised centre, following minor blow to thigh in severely affected haemophilia A boy.



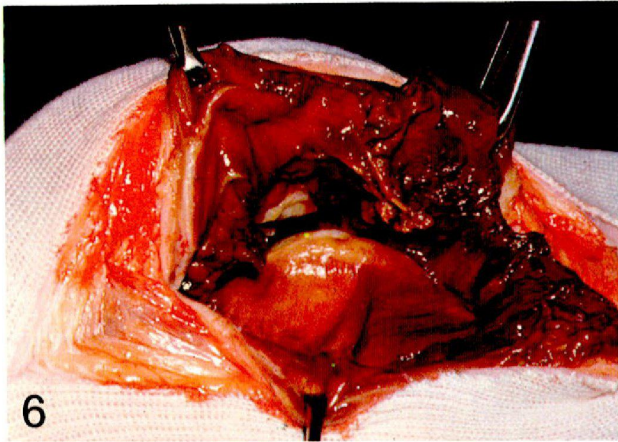
3
Massive intramuscular haemorrhage into thigh, following impact with the corner of a table; a tension blister is present. Haemophilia B patient.



4
Autopsy photograph showing immediate pre-mortem bleed between fibres and sheath of a forearm muscle. Haemophilia A road traffic accident.



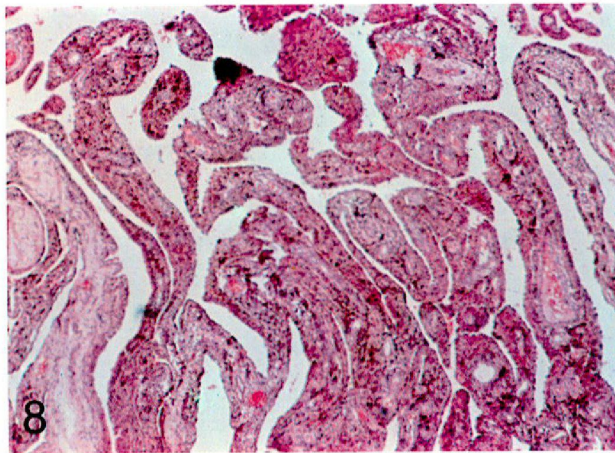
5
Knee opened in mildly affected haemophilia A man (factor VIII 20%), three months after removal of meniscus without replacement therapy cover. Post-operative haemorrhage (the patient's first haemarthrosis) had occurred and had not responded to conservative management. At operation a pseudo-tumour arising from a nick in the tibial cartilage was excised. Plate shows a) bruising deep to the cartilage, b) a superficial erosion and c) pannus being lifted off by forceps. Plate suggests that significant changes may result from first haemarthrosis and illustrates need for early and energetic therapy at the first evidence of joint involvement in childhood.



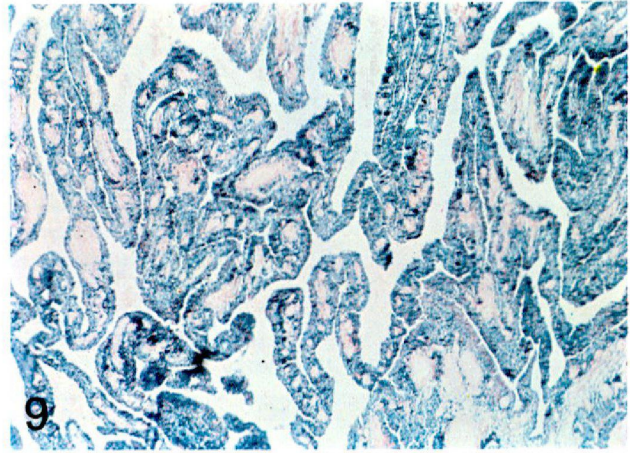
6 Knee, severe haemophilia A. Hypertrophy and staining of synovium.



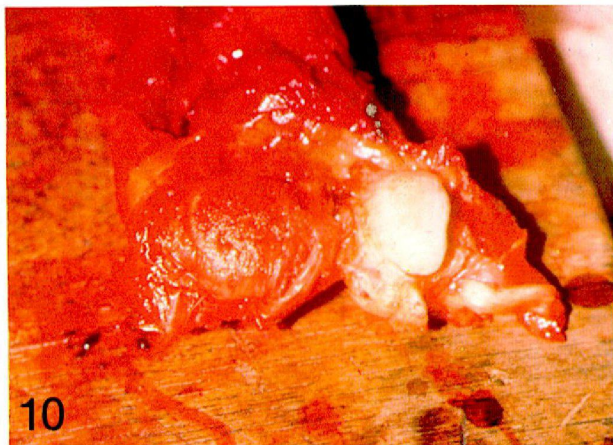
7 Specimen of synovial membrane removed at synovectomy; severe haemophilia A. Specimen suspended in saline, showing hypertrophic villus formation.



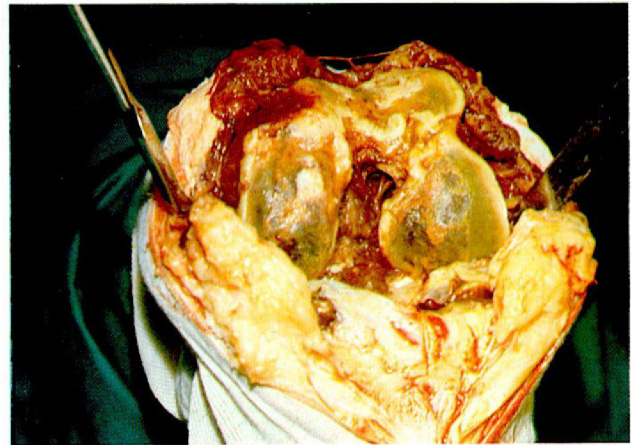
8 Photomicrograph synovium; H and E.



9 Photomicrograph synovium; stained for iron.



10 Lower end humerus. Severe haemophilia A patient at autopsy. Pitting and erosion of cartilage clearly demonstrated.



Chronic haemophilic arthropathy. Lower end femur at arthrodesis. Synovial hypertrophy, hemosiderin deposits and marked erosion of cartilage over condyles, exposing subchondral bone.

CLOSED BLEEDS

Bruising (see colour plates 2 and 3)

Superficial bruising is rarely dangerous and no treatment is indicated provided that

- a) the head and neck are not involved, and
- b) the bruise does not represent a physical sign of bleeding into deep tissues.

Bruising is, of course, one of the hallmarks of a coagulopathy and may be the presenting sign.

In pre-school years most deeper structures are protected by the subcutaneous fat of infancy, but bruising of the forehead, common in the toddler, often presents the dilemma of whether or not to give factor replacement.

It is the author's practice to give one dose of pooled cryoprecipitate (or the appropriate alternative, depending on diagnosis) whenever the head is involved in trauma, but only to admit the patient to hospital for observation and repeated therapy if either the history or physical findings suggest unusual injury.

Other sites of bruising demanding special attention are the neck, abdomen, genitalia and buttocks.

Neck and throat bleeds

These must be treated with immediate factor replacement. Bleeding into the throat may be secondary to infection and the appropriate antibiotic should be prescribed.

Marked swelling of the neck usually indicates a mixture of haematoma and oedema caused by obstruction. Immediate intravenous hydrocortisone (100 mgs) and frusemide (Lasix: 40 mgs) should be prescribed and repeated until the swelling is no longer a threat to respiration. Repeated factor replacement with laboratory control is required.

Tracheotomy should only be employed if these measures fail.

Intracranial haemorrhage

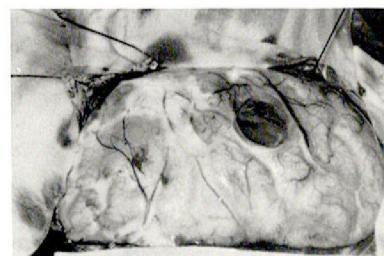
Provided that immediate factor replacement can be given and the haemostatic levels of the relevant clotting factor maintained, the prognosis for intracranial bleeding in haemophilia is the same as for the non-haemophilic population. If possible, blood for antibody screening should be withdrawn before factor replacement is given. Once haemostatic control has been confirmed by laboratory assay, invasive neurological investigation and neurosurgery may proceed.

Antifibrinolytic therapy is indicated.

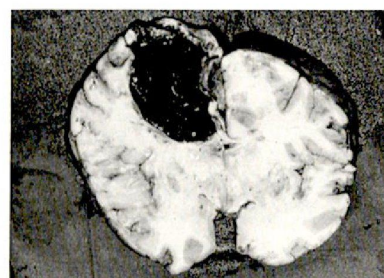
Following invasive investigation or neurosurgery, replacement therapy should be given for a minimum of ten days.

Head injury

Severe blows to the head, with or without skull fracture, should be treated for ten days and the patient should be observed for a further 48 hours after discontinuation of replacement therapy. This procedure should cover the danger of secondary haemorrhage from a minor lesion initially controlled by replacement of the appropriate clotting factor.



Cerebral hemisphere displayed at operation on 11 year old boy with severe factor V deficiency. Subdural haemorrhage at 6 months of age. Note flattening and thinning of cortex with porencephalic cyst. Partial division corpus callosum successfully controlled previously intractable epilepsy.



Intracerebral haemorrhage following road traffic accident. This was a *secondary* haemorrhage and it illustrates the need for full replacement therapy cover for at least ten days after major head injury.

Muscle bleeds

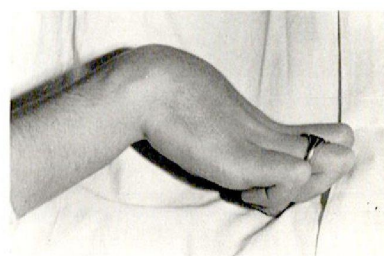
Note: Drugs should be administered by the oral or intravenous route. Intramuscular administration causes iatrogenic intramuscular haemorrhage and its sequelae.

All intramuscular bleeds should be treated with early factor replacement. Bleeds into the following sites, or any intramuscular bleed treated late in its course, require repeated therapy:

forearm bleeds (sequelae: claw hand, Volkmann's ischaemic contracture);

calf bleeds (sequelae: pes cavus);

psoas/iliacus bleeds and retroperitoneal haemorrhage (sequelae: massive blood loss, dehydration, electrolyte disturbances, femoral nerve compression, pleural effusion).



Volkmann's ischaemic contracture in adult with haemophilia A, following untreated forearm bleeds in childhood.

(see colour plate 4)

Forearm and calf haemorrhage require a minimum of 48 hours replacement therapy and subsequent physiotherapy until full function is restored. Light-weight splinting may be indicated.

Psoas/iliacus bleeds

The presentation of this type of bleed is unusual before puberty. It may be precipitated by sexual activity.

Presentation:

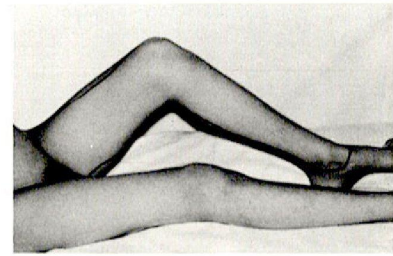
- pain in iliac fossa
- pain on hip extension; patient lies with hip in flexion
- tenderness just below mid inguinal ligament
- tender mass in iliac fossa when presentation is late
- anaesthesia to touch just below inguinal ligament and extending over anterior surface of lower limb in distribution of femoral nerve, extent depending on timing of presentation.

Note: When the bleed is right-sided, presentation may mimic acute appendicitis and its differential diagnoses. Severe loss into the retroperitoneal tissues may be attended by all the physical signs of appendicitis, including foetor oris, vomiting, pyrexia, raised white cell count, abdominal and intrarectal tenderness and rebound tenderness.

The cardinal distinguishing features are the inability of a patient with psoas bleed to extend the hip and the demonstration of anaesthesia over the distribution of the femoral nerve.

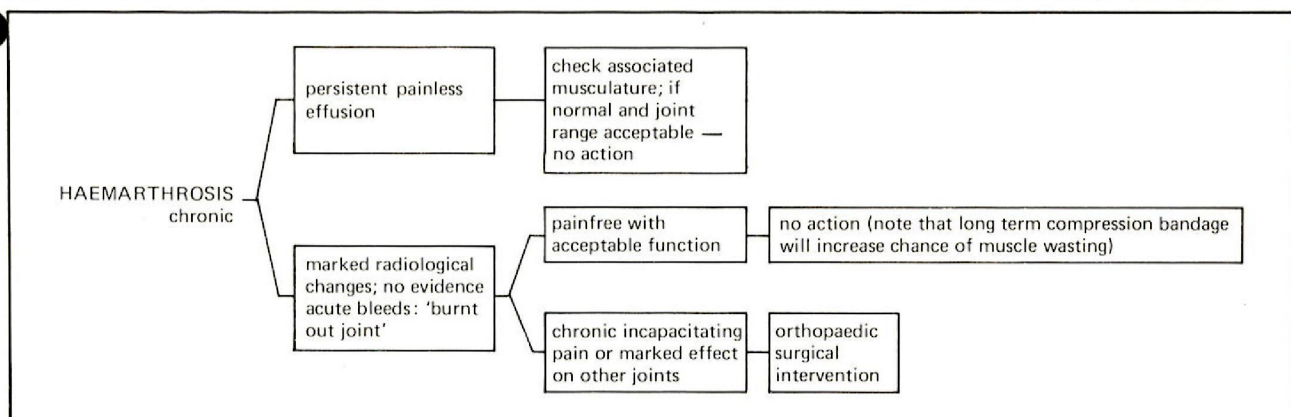
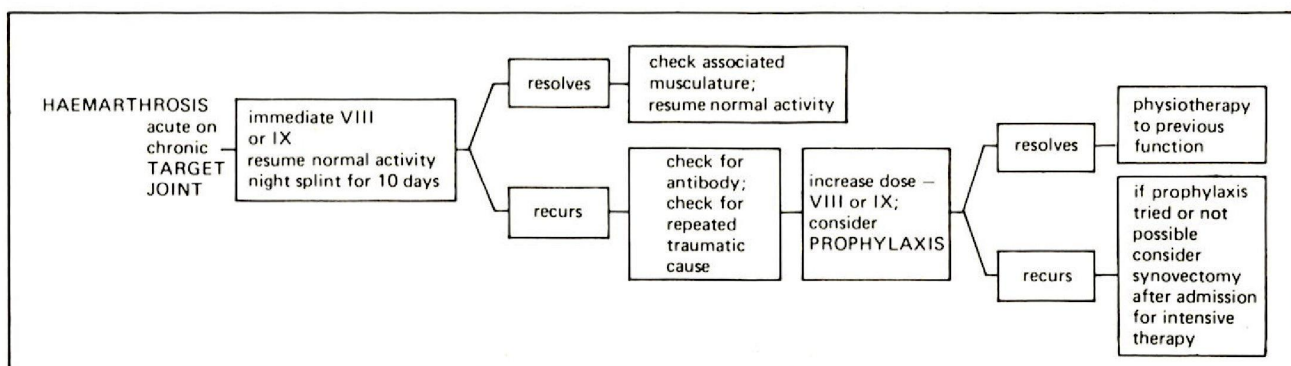
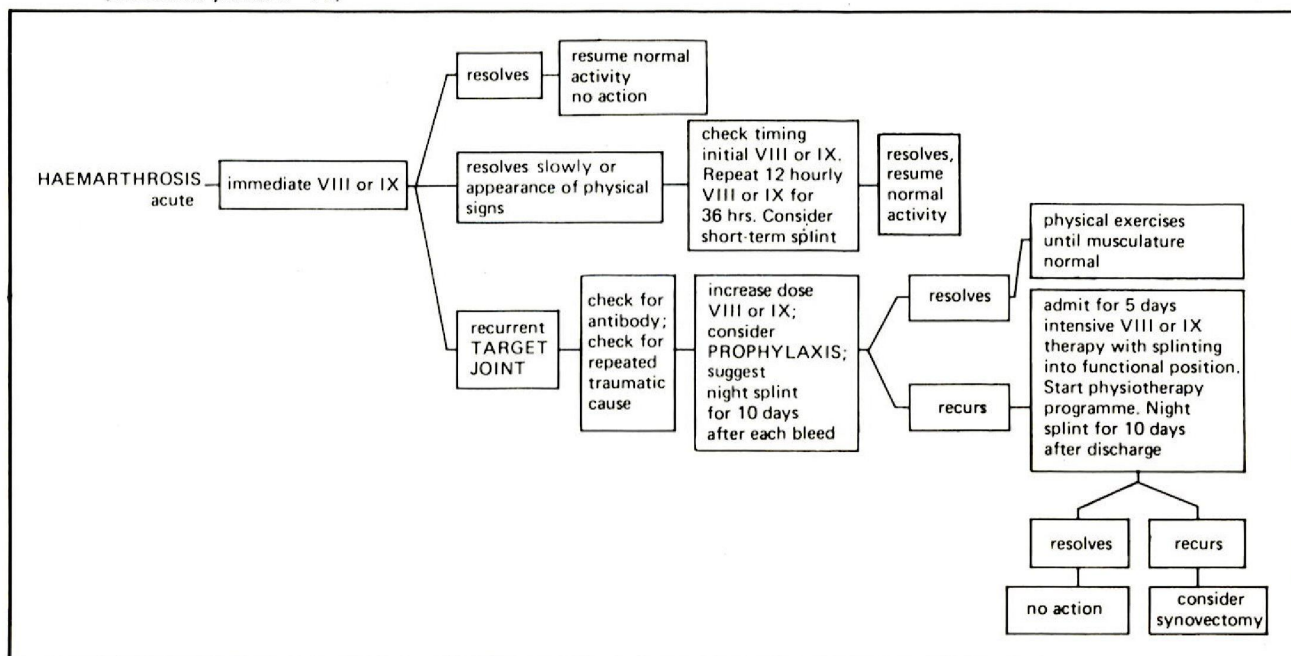
Treatment:

- admission to hospital indicated
- minimum of five days' bed rest
- repeated factor replacement
- fluid balance monitoring
- immobilisation of affected side, when possible and acceptable to patient (with severe haemorrhage, elevation of lower limb in splint — allows flexion at hip and knee — often more comfortable than support with pillows)
- initially physiotherapy contra-indicated (except to other joints and muscle groups). Once bleeding has stopped, cautious physiotherapy required until patient is ambulant and then intensive programme to quadriceps group needed
- abstinence from intense sexual activity advised for one month following bleed.



Position of lower limb following psoas bleed; area of anaesthesia marked.

Haemarthrosis
(see colour plates 5–11)



Treatment with adequate factor replacement is always urgent.

Replacement therapy given as soon as the patient experiences an aura will usually terminate the haemorrhage, allowing immediate return to full activity.

Physical signs of acute haemarthrosis indicate failure of the treatment regime.

Target joints

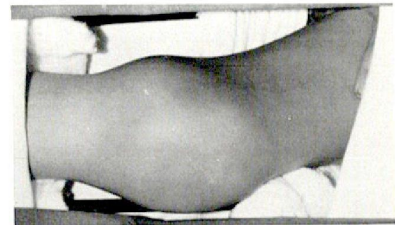
When bleeding into one joint is recurrent, or bleeding occurs into a joint known to be damaged by previous haemarthroses, careful and prolonged supervision with quick and effective VIII/IX replacement and a programme of physiotherapy tailored to the patient are necessary.

Always check for a cause. This is sometimes traumatic and associated with work or a hobby.

Check the gait in the non-bleeding state when recurrent haemarthroses affect the lower limb joints, and X-ray the hips, the knees and the ankles. Abnormalities of bearing or gait can sometimes put a strain on a particular joint, and they are often easily corrected by wedging of shoes, or by light-weight ankle splints which provide support without causing muscle weakness.

Calipers should only be considered as a last resort; muscle weakness and wasting induced by caliper protection are difficult to overcome without the induction of further haemorrhage in the unsupported joint. If calipers have to be used, a regular non-weight-bearing exercise programme should be performed daily on the affected limb.

Flexion deformity or contracture may sometimes be eased out by using serial plaster wedging or reverse traction. In chronic haemophilic arthropathy of the knees, care should be taken to avoid subluxation of the tibia; a 10° loss of extension is usual, and acceptable for adequate function.



Acute knee haemarthrosis. Swelling, and flexion in position of maximum comfort.



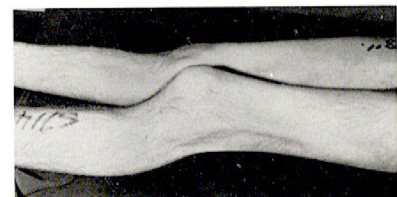
Chronic long-standing haemarthrosis with muscle wasting; severe haemophiliac with high titre antibody.



Long term result of inadequate therapy. Chronic haemophilic arthropathy with marked wasting of quadriceps group and calf musculature.



Very severe chronic haemophilic arthropathy of knee, showing obliteration of joint space, wide intracondylar notch, splaying of bone ends, osteophyte formation and haemophilic cysts.



Subluxation of tibia.

OTHER BLEEDS

Fractures

Long bones: closed, uncomplicated

- routine orthopaedic management
- immediate factor replacement
- once the bleed is immobilised, 48 to 72 hours' further VIII/IX replacement required
- (also *clavicles, ribs, tarsus*)

Long bones: open

- routine orthopaedic management under VIII/IX cover
- once the wound is immobilised, five days' replacement therapy recommended

Facial bones and carpus

- routine orthopaedic management
- duration of factor replacement therapy will depend on extent of damage

Skull, spine, pelvis

- replacement therapy for *minimum* of ten days

Complications

Secondary haemorrhage may occur despite adequate VIII/IX cover, especially when the neck of the femur and the hip are involved.

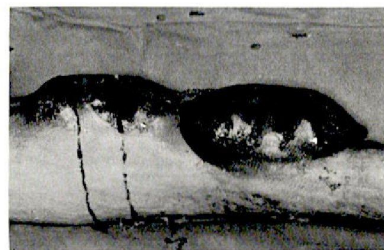
Replacement therapy depends on the extent of the injury or the surgery and the nature of the complication.

Remember tetanus prophylaxis for open wounds.

For VIII/IX dosage, see Booklet 3 on *Therapeutic Agents*.

Pseudotumours

The management depends on the site, extent and disability caused by the pseudotumour. Surgery, often extensive, should only be performed in specialised Centres.



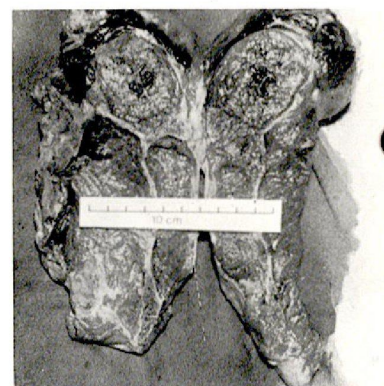
Massive ulcerating pseudotumour of calf. Ten year history of development. Severe haemophilia A.



X-ray of lesion showing bone destruction and calcification of soft tissue.



Post-operative result. Note pes cavus associated with untreated intramuscular calf bleeds in childhood.



Other diseases affect haemophiliacs. Clinical diagnosis: pseudotumour scapula. Autopsy diagnosis: secondary hepatoma scapula with associated haemorrhage.

Menorrhagia

This may occur because of low VIII/IX levels in carriers (and in autosomal recessive disorders and von Willebrand's disease). Control may often be achieved by the trial of oestrogen/progesterone (contraceptive pill) therapy, or with anti-fibrinolytic given as oral Cyklokapron (1 to 1.5 grams three times a day) starting on the first day of menstruation and terminating on the last day of menstruation.

Occasionally dual therapy is required.

Factor replacement is rarely necessary.

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5. SURGERY AND DENTAL EXTRACTIONS

5

SURGERY AND DENTAL EXTRACTIONS

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The surgical management of the haemophilic patient should only be undertaken in a hospital with a staff of suitably trained medical and paramedical personnel, comprehensive laboratory facilities and immediate access to sufficient quality blood products to cover both the operation *and* any complications.

The temptation to operate, even on minor lesions, in any other circumstances should be resisted; results are invariably poor, with increased incidence of secondary haemorrhage, infection, delayed wound healing and scarring, bleeding into other sites, increased blood product consumption, analgesic dependency and other psychological problems, prolonged hospitalisation and death.

When emergency surgery *has* to be performed in another hospital — for instance, after a road traffic accident — arrangements should be made to transfer the haemophilic patient to a specialised Haemophilia Centre with minimum delay.

INDICATIONS FOR SURGERY IN THE HAEMOPHILIAC

Relief of pain and suffering	Example: arthrodesis for the chronic incapacitating pain of severe haemophilic arthropathy.
Relief of recurrent haemorrhage	Example: synovectomy in a joint subject to recurrent bleeding and not responsive to adequate on-demand or prophylactic factor replacement and physiotherapy.
Restoration of function	Example: hip arthroplasty for destruction of joint by haemophilic arthropathy.
Voluntary sterilisation	Example: vasectomy requested by haemophiliac and his wife, to prevent conception of carrier daughters.
General surgical indications	Examples: appendicectomy; removal of tumour; haemorrhoidectomy.

CONTRA-INDICATIONS TO SURGERY IN THE HAEMOPHILIAC

Note: None of these contra-indications is absolute.

The decision to operate depends on the specific requirements in a particular case.

In general, non-surgical management should always be considered as the first choice in any case.

The presence of an anti-body to factor VIII (or other relevant clotting factor)

The presence of a concomitant haemostatic defect

The presence of HB_sAg positivity

The possibility of non-surgical management of the lesion

Examples: thrombocytopenia or qualitative platelet defect induced by drugs; severe liver disease.

Examples: intensive factor replacement for a muscle haematoma; factor prophylaxis for recurrent haemorrhage; cimetidine therapy for peptic ulceration.

PRE-OPERATIVE INVESTIGATION AND MANAGEMENT

Assessments should be made of:

- *general health* — including renal and hepatic function, state of the veins and psychological stability;
- *haemostatic function* — including a review of primary diagnosis and exclusion of a clotting factor antibody.

CHECKLIST

History

- general
- specific, including state of veins
- social, including review of smoking, alcohol, analgesia

Examination

- general
- specific, including musculo-skeletal assessment

Radiology

- chest
- specific

Urinanalysis

- blood, protein, glucose

Blood

- haemoglobin, white count, platelet count
- factor level
- factor antibody screen
- urea
- liver function tests
- HB_sAg/Ab
- blood group; cross-match if necessary.

Instructions for

Nursing staff

Intramuscular injections contra-indicated — give drugs orally or intravenously.

Only implement major changes in management after consultation with haemophilia team.

Physiotherapist

Maintain joint ranges and muscle function.

Implement physiotherapy to operation site only after factor replacement and consultation with haemophilia team.

Anaesthetist

Intramuscular pre-medication contra-indicated.

Note previous analgesia and possible tolerance to specific drugs.

Note previous history liver disease.

Surgeon

Operation should only proceed when factor anti-body screen negative and **post-transfusion assay shows adequate haemostatic level has been achieved.**

ON THE DAY OF OPERATION

Transfuse relevant blood product

Assay pre- (if previous blood product therapy recent) and post-transfusion levels

As there is often an immediate rise followed by a rapid partial fall in the *in vivo* factor level, at least two assays (ten minutes and thirty minutes) are recommended.

If response satisfactory inform surgeon

Give first dose of antifibrinolytic intravenously if indicated

Repeat factor assay at end of operation

Remember that a percentage of the transfused factor will be used up during the operation, and a further dose may be required within a few hours of surgery.

Thereafter, therapy depends on the nature of the wound and on the response and half-life of the transfused factor in the individual patient.

Duration of therapy

Unless complete immobilisation can be guaranteed, replacement therapy should be maintained until wound healing is well advanced; when calculating blood product requirements pre-operatively, allowance should always be made for

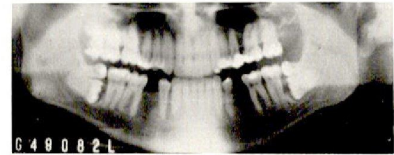
at least three weeks' therapy, with further product to cover mobilisation.

Analgesia

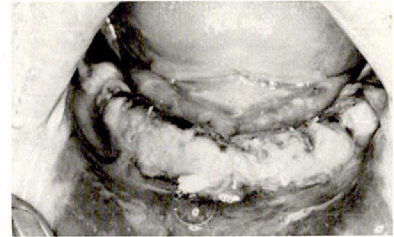
The post-operative analgesic requirement of the severely affected haemophilic patient may be higher than usual because of tolerance. Regular analgesia, rather than an as-required prescription, should be given for the first 48 hours. In pre-operative consultation the patient should always be warned to expect pain at this stage and informed of the measures that will be taken to counteract it.

DENTAL EXTRACTIONS

With the advent of routine antifibrinolytic therapy dental extractions rarely present a major management problem, *provided that an antibody has been excluded.*



X-ray of the teeth of a severely affected haemophiliac, showing both impacted and supernumary lower 8's



These were removed together with the remaining teeth. The photograph was taken on the day after the operation when the patient was discharged from hospital. He was treated according to the recommended procedure and required no further blood product therapy after the day of operation.

Routine varies from Centre to Centre, but the following procedure is well-proven and will serve as a guide to management

- exclude factor antibodies
- check urine (haematuria or evidence of renal dysfunction contra-indicate antifibrinolytic therapy)
- transfuse relevant blood product, calculated to raise factor level to twenty-five percent
- give first dose of antifibrinolytic intravenously
- extract teeth under general anaesthesia (children) or local dental block (adults)
- insert pledget of absorbable material soaked in sterile topical thrombin into sockets
- oversee sockets with catgut mattress stitch
- prescribe antibiotic if indicated
- continue antifibrinolytic orally for one week
- ban harsh foods such as toast, crisps, nuts, etc for one week

When admission to hospital is necessary

day 1 — admission
day 2 — extractions
day 3 — discharge

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6. MANAGEMENT OF FACTOR VIII ANTIBODIES

6

MANAGEMENT OF FACTOR VIII ANTIBODIES

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Seven percent of haemophilia A patients in the United Kingdom develop antibodies. The management of these cases is at present the subject of intensive discussion. Although no definitive policy can therefore be recommended, these guidelines to management may be helpful.

DETECTION OF ANTIBODY FORMATION

Maintain a high level of suspicion

In treating any group of haemophiliacs, the doctor must constantly bear in mind the fact that antibody formation will occur in a proportion of patients.

Severely affected patients should be periodically subjected to an antibody screen, especially when they are on home therapy.

Antibodies may present with a gradual failure of previously effective treatment, or they may be discovered on the routine mandatory check before surgery.

Early recognition

Early recognition of antibody formation may be important, as there is some evidence that immunosuppressive therapy may be effective if it is given during or soon after the primary immune response.

It is not yet possible to predetermine which haemophiliacs are going to develop an antibody.

ASSESSMENT OF RESPONSE TO THERAPY

Laboratory parameters

Allain has suggested that patients with antibodies be divided into two groups, 'high responders' and 'low responders'.

High responder antibody patients show an anamnestic response four to six days after therapy with factor VIII, high titre antibody production precluding further therapy with even very high doses of factor VIII for several months.

Low responder antibody patients may show a measurable antibody response to factor VIII stimulation, but this is of low titre and is unlikely to preclude further therapy with high dose factor VIII.

The difficulty is that these differences are not always clear-cut. Responses vary with time and the same patient can react in different ways.

Clinical effect

Clinical experience suggests that small doses of factor VIII given early in the course of a bleeding episode may stop the bleed despite the presence of an antibody; this may be because the clotting

sequence at the site of the bleed is completed before antigen destruction occurs.

METHODS OF TREATMENT

The choice of treatment will depend on:

- the nature of the lesion;
- its prognosis;
- the antibody titre at the time;
- the previous history of the patient;
- when the antibody developed;
- how the patient has responded to therapy since its development.

These variables and the different therapeutic approaches have been summarised by Penner and others.

Immunosuppression

Haemophilic patients in their first primary response or non-haemophilic patients developing an anti-factor VIII antibody in the course of another disorder (usually 'collagen-vascular') may benefit from immunosuppressive therapy.

Dormandy and Sultan, in an enquiry for the World Federation of Hemophilia, studied reports on fifty-six patients treated in this way, and they concluded that in seven cases immunosuppression might have had a beneficial effect.

Cyclophosphamide was the drug used most frequently in the survey, but azothiaprime has also been employed. The value of steroids is uncertain.

Suggested cyclophosphamide dosage

- 1 Give factor VIII at dose calculated to raise level in patient by 60%.
- 2 Through same drip needle administer i.v. cyclophosphamide 7.5 mg/Kg body weight.
- 3 At 24 hours administer i.v. cyclophosphamide 5.0 mg/Kg body weight.
- 4 At 48 hours administer i.v. cyclophosphamide 2.5 mg/Kg body weight.
- 5 Prescribe *oral* cyclophosphamide 3 mg/Kg body weight for 7 days.

Reference: Dormandy K, personal communication.

Note: Cyclophosphamide may induce acute nausea and vomiting, which can be prevented by the prescription of an antiemetic.

Factor VIII

If elective surgery is contemplated, factor VIII in any form should be *withheld* to allow the antibody titre to return to zero before operation.

This buys four or five days' effective therapy with high dose factor VIII for the patient, and early haemostasis is ensured.

In these cases surgery must be planned to follow the first factor VIII dose immediately.

In severe haemorrhage, suggested factor VIII dosage is 10,000 units twice daily.

As explained under *Clinical effect*, low dose factor VIII therapy may have a place in the management of antibody patients.

**Animal factor
VIII**

Although there is interaction between the human and animal products in terms of antigenicity, the antibody titre may initially be lower to the animal products and a higher, and perhaps longer, response may be possible.

Note: Porcine and bovine AHG are unavailable in many countries.

**Prothrombin
complex
(‘Activated
factor IX’)**

There is now substantial evidence that haemorrhage in haemophilia A patients with antibodies is controlled by some preparations of the prothrombin complex (II, VII, IX, X). The active ingredient of these preparations is thought to be X_a , which bypasses the need for factor VIII in the clotting sequence.

The thrombogenicity of prothrombin complex products has been well documented, and disseminated intravascular coagulation is a consistent feature following injection into animals. It follows that careful monitoring for unwanted side-effects is required when these products are used. Shortening of the whole blood clotting time, or of the prothrombin and activated partial thromboplastin times have been suggested as measures of control, but a standard ‘unit of activity’ has yet to be defined.

As thrombogenicity appears to vary from batch to batch it is not possible to recommend specific products. Either an *in vitro* demonstration of APPT correction when the product is added to factor VIII deficient plasma, or possibly an animal test, should precede clinical use. Dosage is empirical, recommendations being packaged with the product.

Prothrombin complex preparations used in Haemophilia A antibody patients include Auto-IX, Konyne, Fraction R – renamed FEIBA (Factor Eight Inhibitor Bypassing Activity) – and CAP (Controlled Activated Product).

Further investigation of the efficacy and the dangers of the prothrombin complex products is being undertaken by a task force of the International Society for Haemostasis and Thrombosis.

For further information, see ‘Workshop on Inhibitors of factors VIII and IX’ published by Facultas-Verlag, Wien, for Immuno AG, 1976, and the proceedings of the 3rd European Workshop (in press as supplement to Scandinavian Journal of Haematology).

Subsidiary methods

Immobilisation

If a clot can be formed by any of the above methods, it is vital to leave it undisturbed until consolidation can occur and wound healing is complete. Non-adherent dressings should always be used.

Sedation and splinting may be necessary, as may the acceptance of secondary problems such as bedsores, which would normally receive energetic therapy.

Local haemostats

The local application of absorbable material soaked in topical thrombin or pastes made from factor VIII concentrate may be useful in controlling haemorrhage.

Antifibrinolytics

Provided that renal pathology (including haematuria) is excluded antifibrinolytic therapy is worth a trial in the treatment of bleeding in patients with antibodies.

There is some evidence that Cyklokapron increases the effectiveness of factor VIII treatment.

Plasmapheresis

Plasmapheresis using a continuous flow cell separator has superseded earlier attempts to reduce the level of antibody by exchange transfusion. Good venous access is essential. Plasma removed during the procedure should be replaced with a factor VIII-free product, such as plasma protein fraction (PPF). As there is a substantial risk of aerosol contamination in the use of these machines the question of the Hb_sAg status of the patient should be considered.

From the available reports it appears that the technique should certainly be considered in acute situations. There is, however, less convincing evidence of long-term beneficial effects with regular plasmapheresis.

Estimations of antibody level should follow plasmapheresis; occasionally an antibody of higher titre than that measured prior to the procedure has been recorded.

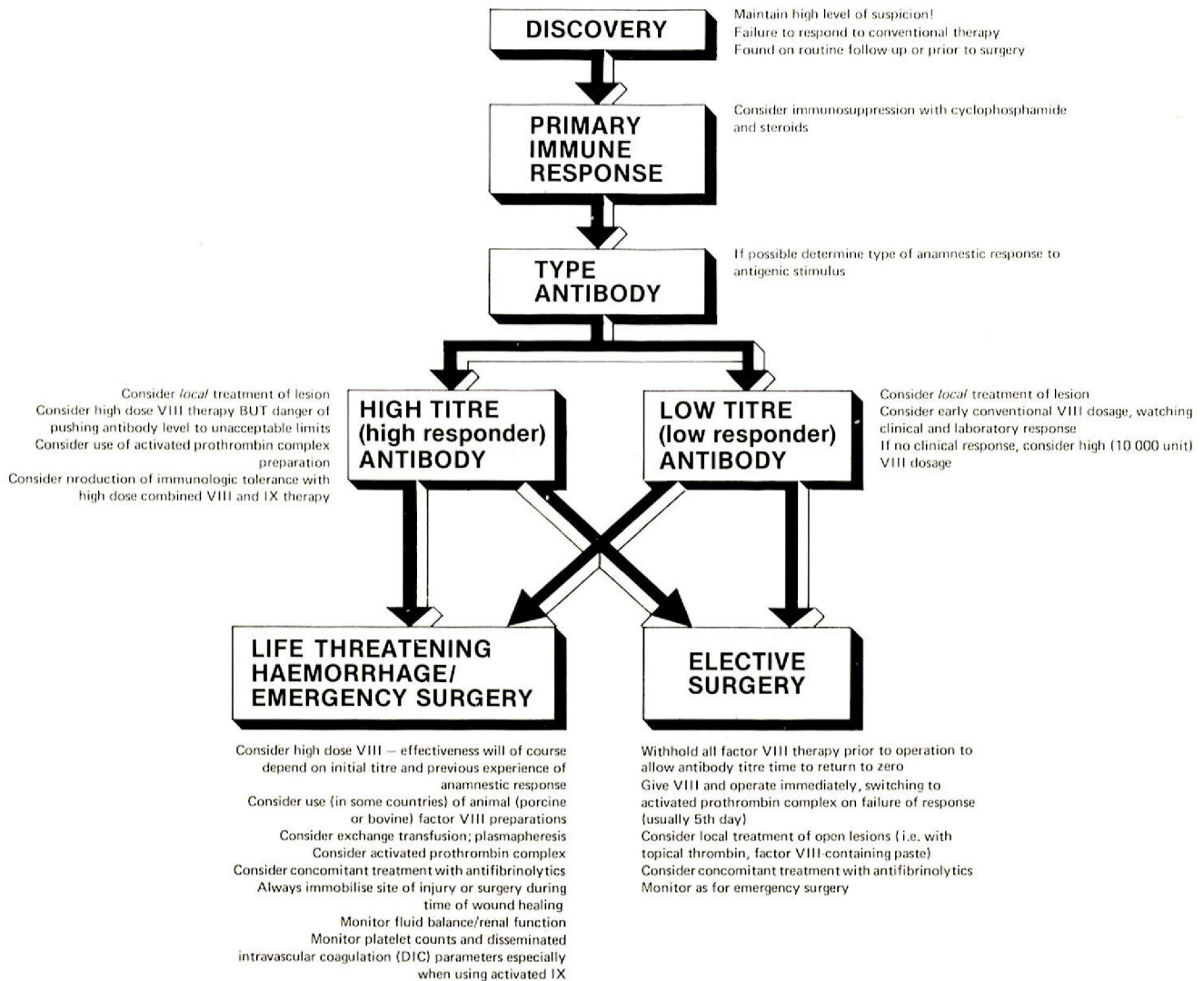
Continuous infusion

Continuous infusion of factor VIII concentrate, or regional perfusion of the site of injury should be considered.

Immunological tolerance

Brackmann and his colleagues have suggested that combined factor VIII and IX therapy in *very high* dosages over periods of many months results in the disappearance of high titre VIII antibodies. It is possible that such an effect may be due to the development of immunological tolerance to factor VIII antigen in the patient.

ANTIBODIES—A GUIDE TO MANAGEMENT



Note: Management of antibodies (VIII inhibitors) remains controversial at the present time and much depends on the individual patient and the nature of his lesion. This scheme is suggested for guidance only.

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7.

SOCIAL MEDICINE

7

SOCIAL MEDICINE

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Starting a haemophilic patient on a home therapy programme, whether this involves self-infusion or training family members to take the initial responsibility for treatment, does not reduce the need for social work provision within the Centre

Although social work intervention may be needed at any time with haemophilia, stress is more likely at certain stages in the life of the affected family

Encouraging independence and teaching patients to take the initiative for their own care changes the relationship between the haemophiliac and his family, and the Centre, but it is vital for this relationship to be maintained.

This booklet explores briefly the scope of social work within the Haemophilia Centre, and is based on the idea that home therapy grows out of a progressive relationship with a family, which eventually matures into a partnership between the haemophiliac and his advisers.

- during courtship and early marriage of the haemophiliac or haemophilia carrier
- during pregnancy and parturition, and in the immediate post-natal period, depending on the sex and diagnosis of the infant
- at positive diagnosis and during the following six months
- during infancy and the toddler stage of a haemophilic boy
- the first time away from home
- in the immediate pre-school or nursery school period
- when an affected boy starts formal education and whenever he changes schools
- at the start of home therapy
- at competitive examinations
- during puberty
- when choosing a career and at interviews
- in early employment
- when on holiday abroad or participating in a new sport or activity which might be construed as risk-taking
- when in hospital for long-term therapy
- when other medical burdens are added to the primary diagnosis of haemorrhagic disorder, e.g. factor antibody, HB_sAg positivity, etc.
- when chronic or acute pain disrupts life or sleep for the patient or his family, or results in concern about analgesic intake
- at expected landmarks in the life of a family, e.g. birth of siblings, bereavements, redundancy of bread-winner, etc.

This booklet provides a simplified checklist of what to look for and what might be done by the social worker and the haemophilia team at each stage

Note: Some elements of each of these stages are considered in the other booklets in this pack, and they are all more fully covered in the work of Alby and Alby, Hurt, and Katz.

Much of the work will be on a one-to-one basis, but consideration should be given to the value of group therapy, either in the hospital, or in meetings organised by voluntary bodies like the Haemophilia Society.

COURTSHIP AND EARLY MARRIAGE

Carrier

The daughter of a haemophiliac, as an obligatory carrier, should be aware of the risks to possible future children before she marries. With her agreement her boyfriend should also be counselled.

It is vital for this counselling to be carried out by experts in the modern management of haemophilia, in private and away from the rest of the family.

The couple should be aware of contraceptive techniques and should have easy access to information on the subject.

Potential carriers should also be offered pre-marital counselling with their intended partners.

Haemophiliac

The haemophiliac should know that *all* his daughters will be carriers and *all* his sons will be normal, and cannot pass on the disorder.

His girlfriend should be encouraged to attend the Centre with him during his routine visits and the couple should be offered joint counselling.

All mature haemophilic boys should be aware of contraceptive techniques, either through instruction by the family, the personal physician or the Centre doctor.

It should be emphasised that haemophilia usually runs true-to-form in a family; any haemophilic grandchildren of a severely affected haemophiliac will be affected to a similar degree.

The social worker has three main roles at this stage

- to *listen* to the worries of each partner
- to *be a source of information*, either directly or by referral to other members of the team.
- to *mediate*, if required, between the couple and their relatives, who may have fears about the wisdom of the marriage.

PREGNANCY AND DIAGNOSIS

The fetus	Parents should know that haemophilia does not increase the risks to the fetus, either during pregnancy or during delivery. Since the clotting factor proteins do not cross the placenta, it is possible to diagnose haemophilia A from a fresh cord blood specimen (see booklet 2 on <i>Diagnosis and Carrier Detection</i>).
Diagnosis with care	A lower than normal factor IX level is common in the newborn, especially in prematurity, and results for haemophilia B should therefore be treated with caution. It is always wise to check the child in the first year <i>whatever the diagnosis</i> .
Normality/ positive diagnosis — informing the parents	<p>Parents should, of course, be told of the <i>normality</i> of a baby <i>immediately</i>, but it is sometimes difficult to be sure that it is right to tell them immediately that their newborn infant has haemophilia. It is probably better to wait for a few days to allow normal bonding to start, having informed the parents beforehand that the laboratory investigation may take several days.</p> <p>When the diagnosis is given, it should be imparted to both parents together, with the baby present. The baby should be examined by the team paediatrician at this time in front of his parents, and his normality, in every respect apart from haemophilia, should be emphasised. The social worker should either be present at this consultation or should see the parents immediately afterwards.</p>
Post-natal support	Parents need a great deal of support in the six months following positive diagnosis. This must be characterised by a positive forward-looking attitude, engendered by the paediatrician at the first examination and fostered by the social worker and the rest of the team.

PRE-SCHOOL

The first signs	<p>The first <i>signs</i> of a bleeding disorder usually appear when the child becomes mobile. It is, of course, at this stage that non-familial cases of severe haemophilia present for diagnosis; the suspicion of non-accidental injury may have been raised with the parents by the time they are seen at the hospital, and they may feel upset and threatened.</p> <p>During the toddler stage parents will be learning which physical signs indicate the need for treatment, but the ability to judge can only be achieved by taking the child frequently to a Centre or to their doctor. A <i>home visit</i> by the social worker should be considered during this period.</p>
Play and punishment	Many of the problems that arise are those normally faced by parents with their first child, but some of them, especially those concerned with

punishment and with play away from home, are heightened for the parents of a haemophiliac.

Parents, especially the mother, often need reassurance that it is not wrong to smack a naughty haemophilic boy, and that it is harmful to keep a child alone at home, rather than to let him play with other children.

**The father's
role**

The father may feel left out of decision-making and play, particularly if a grandparent lives near the nuclear family. He must be encouraged to play an active parental role; his relationship with his haemophilic son is of vital importance at this stage.

HOME THERAPY

Home therapy is not usually introduced until a child is five or six years old. By this time parents will be used to contacting the Centre with their problems, and to frequent visits for follow-up or for the treatment of acute bleeding episodes. Even when the hospital has simply been the place to go to for a transfusion, a sudden change to home therapy can be frightening.

**Getting used
to the idea**

A home therapy programme demands the attention of the social worker as much as that of the nurse, the doctor and the physiotherapist.

The idea that parents should be trained to inject their son intravenously should be introduced gradually from an early age. The thought of venepuncture is repugnant to most people at first, but encouraging parents to participate in the techniques stage by stage always overcomes this fear.

Again, a home visit by the social worker or Centre nurse should be considered. It demonstrates to the family the willingness of the Centre staff to continue to be involved with everyday care after the start of the programme. It also gives the social worker the opportunity to assess the home environment — is it suitable for intravenous techniques, asepsis and proper attention to the disposal of soiled materials?

**Liaison with
the doctor**

Although some doctors like parents or patients to telephone for advice before giving intravenous therapy, this is usually unnecessary. The family must know, however, that they *can* ring and speak to a familiar person if they get into difficulties.

Maintaining contact

A major role of the social worker in a home therapy programme is that of ensuring that contact with the Centre is maintained. Although independence is to be encouraged this should not jeopardise the social, physical and mental health of the patient or his family.



Steep stairs from an upper level apartment are an invitation to injury for the haemophilic child or an adult with haemophilic arthropathy.

EDUCATION AND EMPLOYMENT

Anticipating the problems

Throughout a haemophiliac's education, career guidance and employment the social worker's chief task is to *anticipate*.

With the parent's approval contact is made with the nursery or first compulsory school well before the starting date.

NEWCASTLE HAEMOPHILIA SURVEY EFFECT OF HAEMOPHILIA ON EDUCATION 84 haemophiliacs who had completed formal education

Very bad	23 (27%)
Bad	11 (13%)
Moderate	16 (19%)
Little effect	15 (18%)
No effect	19 (23%)

The combined impact of loss of schooling and resistance by employers to haemophiliacs, especially in a climate of high unemployment.

NEWCASTLE HAEMOPHILIA SURVEY UNEMPLOYMENT 1973-74

UK	Region	Severe haemophilia
2.9%	5.1%	33%

When necessary, visits should be made and teachers seen personally; if a school health service exists communication may have to be through its officials rather than directly with the school.

Again with the parents' approval, copies of school reports should be seen by the Centre and any real problems referred to the clinical psychologist.

Career guidance should be sought in good time, and every effort should be made to ensure that every haemophilic boy leaves school with the real prospect of a job or further education.

Arrangements for prophylactic therapy might be made to cover competitive examinations and interviews.

Informing employers

The decision to inform prospective employers of the bleeding disorder *lies with the haemophiliac himself*, and he should make his decision in the knowledge that the Centre staff will respect it, and that they are willing to see the employer personally if he requests it.

Many employers are under the impression that haemophiliacs exsanguinate themselves after any minor injury. They should be assured that *normal first aid methods apply*, and that haemophiliacs bleed no faster than anyone else. They should also know that there are different severities of haemophilia, and that the disorder does not preclude manual work.

PUBERTY

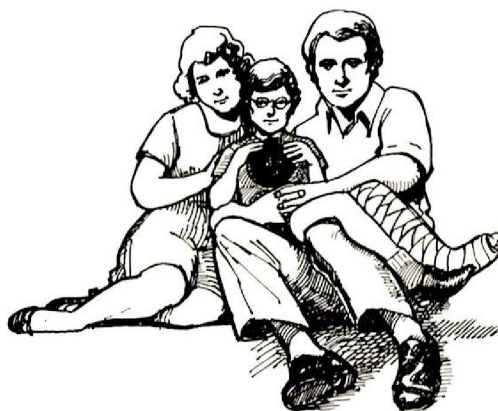
Coming to terms

If he has not already done so, puberty is the time when the haemophilic lad may rebel against his disorder before learning to live with it. It is at this age particularly that his father can be of help, especially if most of the haemophilia team is female. The social worker's task is to explain to the family the boy's natural fears about his haemophilia which have been superimposed on the normal stresses of growing up.

ACTIVITIES AND HOBBIES

Overprotection

It is perhaps in this area that the social worker can do most to guide the family away from the temptation of overprotection, and to encourage them to allow the haemophilic boy to develop normally. Rule-of-thumb advice is to let the child do anything his siblings or friends are doing and to let him learn from experience.



The overprotective attitude represented by this family scene *must be avoided*. The haemophiliac must be allowed and encouraged to develop normally.

The only bans that should be suggested are body-contact sports like boxing or rugby or American football, which may result in head injury.

Holidays

Holidays away from home should be encouraged, as long as cover is provided by home therapy training, the provision of a holiday kit and a letter of introduction to the nearest recognised Haemophilia Centre. (See: Abbott Guide for Travelling haemophiliacs; National Directory, USA)



Haemophiliacs on a sporting holiday organised by the Pisa Haemophilia Centre. Swimming is a particularly good sport for haemophilic boys.



Haemophiliacs should be encouraged to take part in sporting activities such as running and gymnastics.

CHRONIC ILLNESS

Teamwork Long-term hospitalisation, chronic painful arthropathy or the presence of another disabling disorder, in either the patient or his family, demand skilled social support. Families with chronic incurable long-term problems may present an individual social worker with too great or too time-consuming a task, and the load should be shared wherever possible. In situations like this the possibility of short-term respites for other members of the family, by providing institutional accommodation, should be borne in mind.

Awareness The social worker and the rest of the team should be aware of problems which at first sight may seem insignificant; a family may, for instance, have difficulty in financing travel to and from the hospital. This should be anticipated and help offered. The patient's family can well do without this kind of additional worry.

At all stages the Social Worker should be available as a source of information on benefits and services to patients and their families.

CHECKLIST

It is suggested that
these facts
about the
patient should
be known by
the Centre

- | | |
|------------------------------------|---|
| Personal | <ul style="list-style-type: none">— age— nationality— religion— next of kin— marital status— details of family — names, ages and occupations of parents, siblings, children— family history bleeding disorder— degree of disability/clinical severity/antibody— presence of disorder other than haemophilia |
| Housing
and environment | <ul style="list-style-type: none">— type and ownership of accommodation— suitability for severity of haemophilia and need for modification or rehousing— existence of garden/suitable play-area for children— ease of access to local amenities |
| Mobility | <ul style="list-style-type: none">— transport used by family— transport to school/work— provision of wheelchair— ambulance arrangements |
| Financial | <ul style="list-style-type: none">— standard of living— social security benefits— insurance |
| General | <ul style="list-style-type: none">— tobacco/alcohol consumption— hobbies/activities/sports— bans placed on activities by family/doctor— membership of voluntary societies, especially Haemophilia Society— holidays |
| Education | <ul style="list-style-type: none">— where/when/how much lost because of haemophilia— qualifications and attainments— higher education— patient's goal |
| Employment | <ul style="list-style-type: none">— registered disabled?— employment history— type and suitability of job— does employer know of haemophilia?— socio-economic class |
| Treatment | <ul style="list-style-type: none">— attitude to treatment of haemophilia— suitability for home therapy— understanding of reasons for treatment of specific bleeds— fears about treatment— attitude to regular follow-up— attitude to regular exercise |
| Family planning | <ul style="list-style-type: none">— understanding of inheritance patterns— desire for children— contraception/sterilisation |
| Pain | <ul style="list-style-type: none">— type — acute/chronic— analgesic used— degree of interference with life/sleep/work |

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8.

HOME THERAPY

8

HOME THERAPY

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The term 'Home Therapy' includes intravenous treatment given at work or at school as well as at home. The treatment might be self-administered by the haemophiliac or it might be given by a suitably trained relative or friend. When a full home therapy regime is not justified, for instance when bleeding is infrequent, and the patient's home is a long way from the nearest Haemophilia Centre or hospital, a family doctor is often willing to administer or supervise injections.

Home therapy covers both on-demand or crisis therapy, that is injections given on the first evidence of bleeding, and also prophylactic therapy.

Although it is generally agreed that the lyophilised concentrates are the best products for home therapy, cryoprecipitate (or, in the case of factor V deficiency, for example, fresh frozen plasma) might be used.

Many of the firms supplying lyophilised concentrate market home therapy kits which contain all the equipment required for self-infusion. Cryoprecipitate is usually pooled before use, the individual packs being flushed with sterile saline to increase the yield; several devices have been designed to make this task easier and the product therefore more likely to be aseptic.

**The selection
of patients for
home therapy**

Four basic criteria apply to most home therapy programmes.

- The haemophiliac should either bleed frequently enough to justify the training and expense involved, or be isolated geographically from a suitable hospital or doctor.
- In general he should be over 5 years old, have suitable veins and have a relative or friend who is capable of understanding and performing the technical procedures required and of reacting sensibly in an emergency.
- He should live in a reasonable environment and have ready access to a refrigerator for the storage of concentrate (or deepfreeze for cryoprecipitate), and a telephone.
- The haemophiliac or his relative or friend should agree to keep accurate records of all bleeding episodes and treatments and to attend regularly for follow-up.

Recommendations on home therapy criteria were made by a group of interested workers at the First European Workshop on Home Treatment in 1976:

RECOMMENDATIONS ON HOME THERAPY TREATMENT OF HEMOPHILIAS

Selection criteria for patients

A

Candidates should be patients suffering from an established hemorrhagic diathesis, either moderate or severe Haemophilia A or B, or von Willebrand's disease, and who have a need for substitution therapy more than once a year.

B

All candidates for home therapy must be studied for inhibitors. To the present state of our knowledge, patients with high titer inhibitors and those who show anamnestic responses are not eligible for home therapy. In terms of data handling, any patient with an inhibitor should not be included in the home therapy treatment group.

C

If indicated, plasma can be used for home treatment.

D

Minimum age

- 1 For injection by parents: age at which parents can enter the vein and work together with their child (approximately age three).
- 2 For self-injection: age at which the child can find his own veins, can handle the technique and has the emotional stability, maturity and intelligence to manage his own therapy. Whenever possible, training should be initiated prior to puberty.

E

Degree of education, maturity and emotional stability: candidate patients and parents for home therapy should have been assessed as to intelligence, emotional stability, maturity, education and understanding of their illness. It is recognized that the desire to go on home therapy is a very important factor, though not a criterion for exclusion.

F

Social and economic factors: of both the patient and the parents must be assessed and, whenever possible, the assessment must include a home visit. There must be access to a rapid means of communication with the Center. In settings where the cost of hemophilia care is not automatically borne by government or other sources, ongoing attention should be paid to sources of funding of health care.

G

Local physician relations: the extreme importance is recognized of maintaining close liaison between the Hemophilia Center and the local physician, who is providing the day-to-day support of the patient. Wherever possible, this should include a written report of the results of periodic evaluations. The Center ought to be as easily accessible to the physician as it is to the patient.

In 1978 the Haemophilia Centre Directors of the United Kingdom reported criteria for home therapy need in their own Centres:

Criteria for home therapy (51 centres)	No of centres adopting criteria
Severe clinical disease with frequent bleeds	35
Sufficient intelligence to follow instructions, recognise treatment criteria, maintain records, and co-operate with Centre	31
Good veins	23
Psychological stability, reliability, and good sense	20
Ability to perform atraumatic venepuncture	17
Motivation and willingness to give injections	16
Distance from Centre	16
Absence of clotting factor antibodies	14
Good home environment	15
Age	7
Prevention of joint damage	3
Agreement of family doctor	3
Absence of allergic reactions to blood products	1
Absence of hepatitis B antigen	1
Absence of alcoholism or drug abuse	1

The training of patients and families

The criteria to be considered in formulating a training programme were also considered by the European group:

Training program

A

It is the feeling that emphasis should be placed on individual training in terms of the clinic director and his staff teaching one family at a time; this may be well supplemented by group efforts of various kinds.

B

The patient should be given as much theoretical information about his illness as he can handle. This should include insight into antihemophilic factors, the side-effects of the products, the body functions, including joint and muscle. The patient must be brought to understand the importance of earliest therapy. He must be taught that home therapy does not end the hemophilia problems and that all the other supporting measures are still required, with strong emphasis on physical exercise.

C

Practical training should include appropriate sterile manipulations, reconstitution of material, correct venepuncture techniques, attention to disposal of used materials, with a special eye to the prevention and spread of hepatitis. A provision shall be made for adequate storage and inventory of products, and reporting of side effects.

D

The patient should be strongly encouraged to rehabilitation and prevention through physical activity and training.

E

The actual home therapy program should be preceded by an Observation Period of close monitoring.

F

Periodic follow-up of the patient should be mandatory in case of therapy not administered by the patient, and highly desirable in case of self-injection.

G

The benefits are recognized of applying the above training measures to patients who are not candidates for home therapy.

Emphasis is placed on a sound background knowledge of haemophilia and an appreciation of the concepts of good health including regular exercise, as well as on the technicalities of the procedures involved.

Families invited to start home therapy are usually well known to the doctor or Centre staff already and are also usually well versed in the need to recognise and treat bleeds at the earliest possible opportunity. Given this as a foundation, teaching of the actual techniques only takes a day or two. Many children do not like performing venepuncture for some years, and they must not be forced; their parents undertake to perform this part of the procedure.

Some Centres encourage group training sessions in which parents and haemophiliacs discuss all aspects of home and hospital therapy, thereby learning from one another. Such an approach is particularly appropriate during summer camps or at Haemophilia Society meetings.

The treatment plan

Suggestions for the formulation of a set of rules for home therapy were also set out by the Home Treatment Workshop:

Treatment plan

A

It should not be the general rule, after the Observation Period, that the patient must call before infusion for routine hemorrhage.

B

It is left to the discretion of the director of the program to define the pre-set dose and the lesions for which patients must first seek consultation.

C

As appropriate, centers should stand ready to assay in vivo recovery and determine, in multiple samples, the rate of disappearance of the infused materials. One or several centers may decide on adopting fixed sampling time(s).

D

It is recognised that, from time to time, there are patients, both hemophiliacs A and B, who are candidates for prophylactic (continuous) therapy.

E

Appropriate conservative measures should be applied so that, when the patient fails to respond to the therapy, attention be paid to certain aspects, e.g., an inhibitor, an orthopedic lesion, etc.

F

Ancillary therapy should be committed to in a formal written treatment plan, at least one a year, and should include orthopedic, dental, physiotherapeutic, rehabilitative, psychiatric and social care.

Several organisations and Centres have their own rule books and agreed procedures. An example is the text drawn up by the United Kingdom Haemophilia Centre Directors. The following text has been adapted from this booklet.

THE UNITED KINGDOM
HAEMOPHILIA CENTRE DIRECTORS

A HANDBOOK FOR HOME THERAPY

GENERAL GUIDELINES FOR ON-DEMAND THERAPY

<i>The earlier, the better</i>	<i>Early treatment of a bleed prevents later damage. The more blood that is allowed to enter a joint or muscle, the greater the subsequent damage to the tissues and the longer the time taken for recovery. Early treatment usually allows an immediate return to school or work, and it also diminishes the chance of arthritis and disability later in life.</i>
<i>If in doubt, treat</i>	<i>Trust your or your child's aura. If you feel that a bleed might have started, treat it. Never wait until a joint is hot, swollen and painful. Don't worry about 'wasting' the occasional treatment by injecting when a bleed is not present.</i>
<i>A shot in time saves VIII (or IX)</i>	<i>In general, early treatment saves blood product. A small dose of factor VIII or IX stops small early bleeds. Bleeds left to develop require more, and often repeated, doses of blood product to stop them.</i>

WHEN TO TREAT YOURSELF OR YOUR CHILD

<i>An injection of concentrate should be given in the event of:</i>	<ul style="list-style-type: none">— a bleed into a joint— a bleed into a muscle— an injury which you know from experience is likely to result in a bleed
	<i>If you think a bleed is starting, don't hesitate to treat.</i>
	<i>Bleeds into joints and muscles which either don't settle down after two treatments, or which progress in spite of treatment, should be seen at your Haemophilia Centre without further delay.</i>
<i>Immediate treatment should also be given and your Haemophilia Centre notified without delay in the event of:</i>	<ul style="list-style-type: none">— injuries to the mouth, tongue, face, eyes or neck;— severe blows to the head or severe or persistent headache;

- severe or rapid swelling in any site;
- severe pain in the chest or abdomen;
- vomiting, coughing up of blood or the passage of blood in the stools or urine;
- open wounds requiring stitches.

If in doubt, contact your Centre.

Preventing a bleed

There will be times when you want to try to prevent a bleed, for instance before an important examination, an interview or a social event. Your Haemophilia Centre doctor will advise you on the timing and doses of concentrate to give on these occasions.

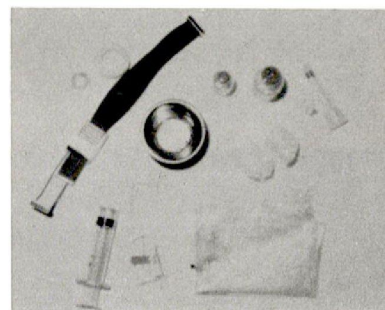
EQUIPMENT

What you will need at home:

- a refrigerator for storage of concentrate or a deep freeze for storage of cryoprecipitate;
- a clean, cool, lockable cupboard for the storage of injection equipment and drugs. This must be out of the reach of young children;
- a telephone.

Equipment checklist

- Cryoprecipitate*
- Freeze-dried concentrate*
- Diluent (sterile distilled water for injection)*
- Disposable syringes: 50 ml, 20 ml, 10 ml and 2 ml*
- Disposable infusion sets*
- Disposable needles*
- Disposable filter needles*
- Disposable butterfly needles*
- Needle disposal box(es)*
- Polythene disposal bags*
- Antiseptic solution (or alcohol-swabs)*
- Adhesive tape*
- Paper towels*
- Polythene sheets*
- Adhesive dressings*
- Tourniquet*
- Thermometer*
- Record-keeping sheets, cards or book*
- Drugs:*
- Additional equipment:*



Equipment required for home therapy.

Note: This list is comprehensive. Inappropriate items should be deleted by the Centre.

HOME TREATMENT PROCEDURE

Preparation for injection

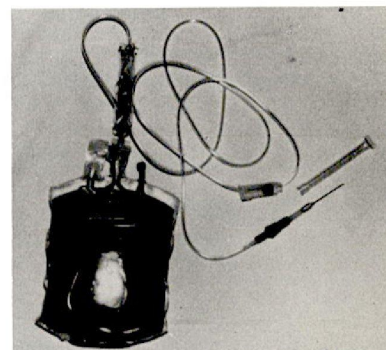
- 1 Prepare a working surface away from food, and cover it with a polythene sheet.
- 2 Wash your hands thoroughly before you begin preparation.
- 3 Collect and lay out all items needed for injection.
- 4 Make sure that a responsible person is with you before you start the injection, and that the person knows the telephone number to ring in the event of an emergency.
- 5 Find a comfortable place to perform the injection, using a covered pillow to support the arm.

Method of mixing and injecting the concentrate

Note: Instructions for the use of cryoprecipitate will be given to you by your Centre Director.

All of the equipment you use is sterile until the covers or containers are opened. Once they are opened, don't touch the needles and don't put them down again without replacing the needle guards.

- 1 Fill a bowl with water, using a thermometer to measure the temperature atdegrees centigrade.
- 2 Remove the protective caps from the bottles containing concentrate and diluent and wipe the rubber stoppers with an antiseptic-soaked swab.
- 3 Attach a plain needle to a.....ml syringe and pull the plunger back untilml of air are drawn into the syringe.
- 4 Pierce the rubber stopper of the diluent bottle, inject the.....ml of air and turn the bottle and syringe upside-down to enable you to withdraw all the fluid.
- 5 Withdraw the syringe and the needle from that bottle, making sure you replace the needle guard before you put the needle down.
- 6 Insert the needle and syringe into the concentrate bottle and gently push all the fluid into the powder. (Some concentrate bottles contain a vacuum.)
- 7 Remove the syringe and discard the plain needle. Then attach a filter needle to the syringe.
- 8 If the concentrate you are using requires it, shake the bottle vigorously for five seconds.



Pooled cryoprecipitate and giving set with protective guard removed.



Cryoprecipitate transfusion via cubital fossa vein. Limb supported by pillow. Drip regulator within easy reach of patient.



After removing protective covers swab tops of all bottles with antiseptic before inserting needle. Do not touch with fingers.

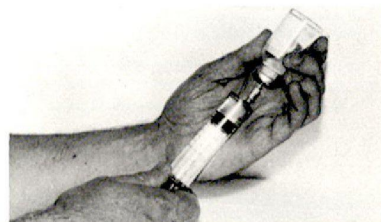


With concentrate bottle on firm surface inject diluent through plain needle.

9 Gently rotate the bottle of concentrate in warm water until all the powder has dissolved. Dry the outside of the bottle.

10 Wipe the bottle top again with an antiseptic-soaked swab.

11 Pierce the rubber stopper of the concentrate bottle with the filter needle, turn the bottle upside-down and withdraw all the fluid into the syringe. Remove the needle from the bottle and replace the needle guard.

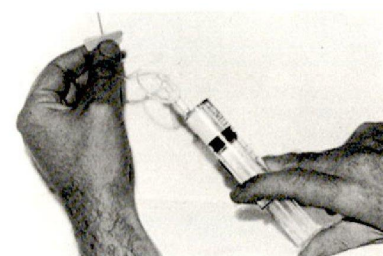


Once all concentrate dissolved withdraw it from bottle using filter needle.

12 Remove the filter needle from the syringe and attach a butterfly needle to the syringe.

13 Holding the syringe nozzle upwards, expel any air by gently pushing the plunger up. Continue until the fluid reaches the end of the needle. Keep the needle guard in position.

14 Sit in a comfortable position with all the equipment within easy reach.



After attaching butterfly needle, hold loaded syringe hub uppermost to expel air. Carefully fill tubing of butterfly set with fluid — bead will appear at point of needle. Needle guard still in position.

15 Apply the tourniquet about 5 cm (2 inches) above the vein to be used and then swab the skin area over and around the vein with anti-septic.

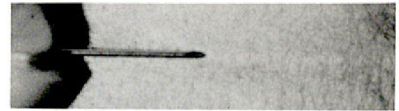


Veins of cubital fossa. Elbow extended. Tourniquet applied.

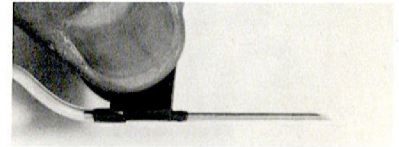


Veins of dorsum of hand. Tourniquet applied. Flexion at wrist and of fingers aids fixation of veins for venepuncture.

16 Remove the protective cap from the needle and then, holding the folded wings of the butterfly, guide the needle through the skin and into the vein. Keeping the skin taut will prevent the skin from moving.

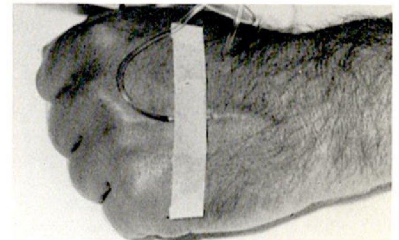


Selected vein on dorsum hand. After anti-septic swabbing line up needle ready for insertion holding folded wings of butterfly firmly.



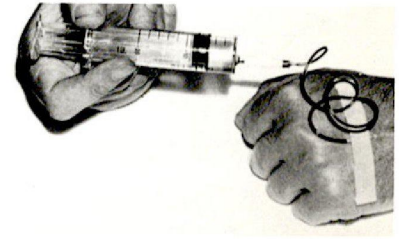
Ensure bevel of needle uppermost. Needle angled at about 10° to pierce skin.

17 Secure the needle in position by sticking a piece of tape across the opened wings of the butterfly. If the needle is in the vein there will be a flow of blood down the tubing.



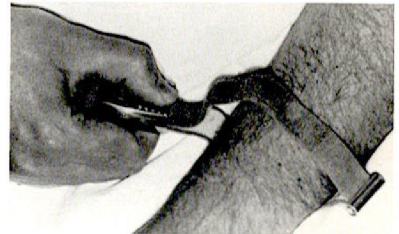
Needle in position — at least 1 cm lies in vein lumen. Successful entry into vein confirmed by appearance of blood in tubing. Butterfly wings open and taped. Ends of tape turned under to allow easy removal.

Note: Don't allow any blood to enter the syringe, because it would clot and the concentrate would then have to be discarded.



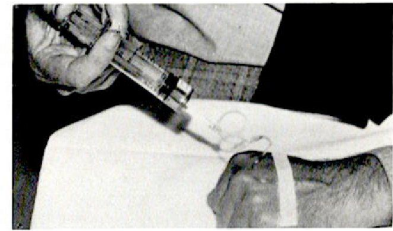
Do not allow blood to flow back into the syringe — clotting will occur. If this happens, discard concentrate.

18 Release the tourniquet.



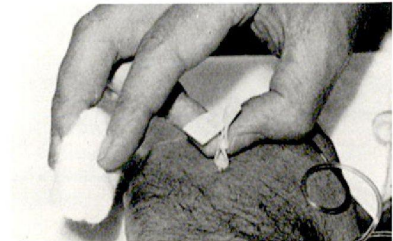
Release tourniquet before injecting.

19 Slowly push the plunger of the syringe until all the fluid is injected, taking three minutes for each 10 ml of fluid. Look at the puncture site; any swelling in this area indicates that the needle is not in the vein. If re-adjustment of the needle doesn't allow correct flow into the vein, remove the needle, press on the venepuncture site for five minutes and repeat the procedure on an alternative vein, using a new butterfly needle each time.

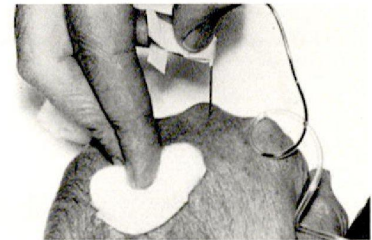


Inject slowly and smoothly — hold syringe hub down so that any remaining air bubbles float to surface of fluid in syringe, and are not injected.

20 When all the fluid has been injected withdraw the needle and immediately apply pressure with a dry swab for at least five minutes. Seal off the puncture site with an adhesive dressing.



Prepare to apply pressure to injection site after withdrawal of needle — hold swab between index and middle fingers, grip folded wings of butterfly between thumb and ring finger.



Immediately needle withdrawn press swab down on venepuncture site.



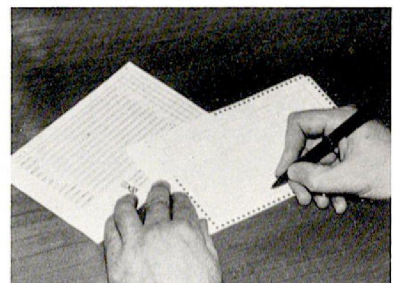
Always cover venepuncture site for at least twelve hours.

21 If a reaction should occur, stop injecting. If the reaction doesn't subside, give as instructed by your Centre Director.

Recording treatment

Make a record of every treatment immediately afterwards. Each treatment must be recorded separately and accurately. All the details must be entered before you go to your Centre for new supplies.

You will only receive new supplies if your records are completed.



Fill in record of treatment carefully.

Storage of equipment

Freeze-dried concentrate (bottles of powder only) are stored in the fridge, on a separate shelf from food, and not in the freezer compartment.

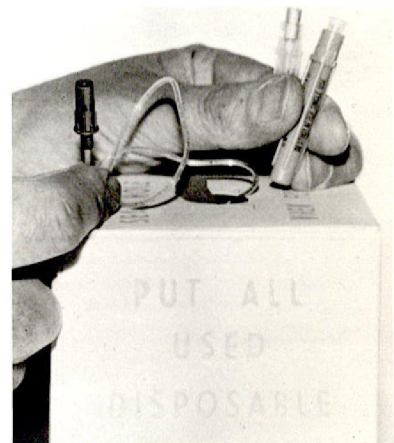
Cryoprecipitate is stored in a sealed container in a deep-freeze, as instructed by your Centre.

All the other equipment must be kept in a separate locked cupboard out of the reach of children.

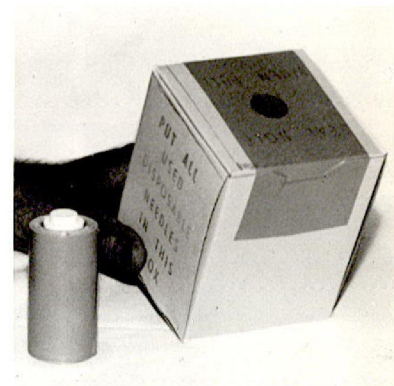
Note: Concentrate can be kept in a cool dark cupboard for two to three weeks (or, only if specified by the manufacturer, for longer periods) or packed in a case for travelling.

Disposal of equipment

All used needles must be put into a needle disposal box. The box should be sealed when it is full and returned to the Centre, or disposed of according to instructions you have been given.



Cover used needles with protective guards and place in disposal box provided.



Seal needle disposal box when full.

All other used equipment must be put into the polythene bags supplied, immediately after use and returned to the Centre for special incineration.

Don't put any rubbish into the waste bins at home.

Special arrangements for disposal may have been made by your Centre Director.

Note: There is always a risk of the hepatitis virus being present in blood products, and so any of the materials you have used could be contaminated. Washing the materials does not get rid of the virus, and so don't allow anyone to play with or re-use any of your equipment.

If any concentrate or blood is spilled, the area should be wiped clean with domestic bleach.

Allergic and other reactions

Reactions to the blood product you are using are extremely rare. You have, however, been supplied with intravenous for such an emergency.

In the event of a reaction you must contact your doctor immediately and not inject any more concentrate until you have done so.

Always record and report any type of reaction.

Never inject anything other than the blood product or the intravenous medicines prescribed by your doctor.

Always use the diluent provided.

If you make a mistake and think that a bottle of concentrate has been contaminated or contains a blood clot, don't use it. Return it to your Centre with your records.

Always use fresh diluent. Never use water left over in a bottle from a previous treatment.

If anyone other than the person being treated is pricked with a used needle, report the incident to your Haemophilia Centre.

CARE OF THE VEINS

Always take your time when you are inserting the needle into the vein. Make sure that everything you are going to need is within easy reach and make yourself comfortable.

During your training you will have been taught which veins are the best and easiest to use. If you treat yourself with care, the same vein can be used for injection every time.

If a vein becomes sore or looks inflamed it should not be used again until it has recovered. If the symptoms persist they should be reported to your doctor.



Place used swabs, towel, polythene sheet, used bottles and syringe in disposal bag and seal.

- The rules of vein care*
- *Always cleanse the skin over and around the vein thoroughly with antiseptic before injection.*
 - *Only inject with new butterfly needles which have not been allowed to come into contact with anything other than their protective guards.*
 - *Always insert the needle into the skin with the bevel of the needle upwards. Use a firm controlled movement at a point which will allow the wings of the butterfly to lie flat on the skin when the needle is in its final position. Keeping the needle in line with the vein and at an acute angle to the surface of the skin, slide it into and up the vein for a short distance to prevent it from slipping out during injection.*
 - *If pain or swelling is experienced at the end of the needle during injection, stop injecting — the point of the needle has either slipped out of the vein or gone right through it. Gentle manipulation will sometimes bring the point back into the vein, but usually a new injection site will have to be chosen.*
 - *Don't apply pressure to the injection site until you have completely withdrawn the needle; pressing the vein wall tightly to the needle may cause damage to the inner lining of the vein. Apply the pressure with cotton wool immediately, as soon as the needle has been completely withdrawn, and keep pressing for at least two minutes, preferably five. Then inspect the venepuncture site and if there is bruising or bleeding apply pressure for a further two or three minutes. Then apply an adhesive dressing and keep it on for at least twelve hours.*
- Remember* *Always release the tourniquet before injecting or, in the event of a failed venepuncture, before withdrawing the needle.*

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9. FOLLOW-UP PROCEDURES AND RECORDS

9

FOLLOW-UP PROCEDURES AND RECORDS

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Regular assessments

Every person with haemophilia should be given the opportunity of attending a clinic for regular assessment, unrelated to his bleeding episodes. It is recommended that severely affected children are examined every six months, and severely affected adults once a year.

Continuity

Apart from these regular, formal reviews of progress, there should be an arrangement for the follow-up of specific problems.

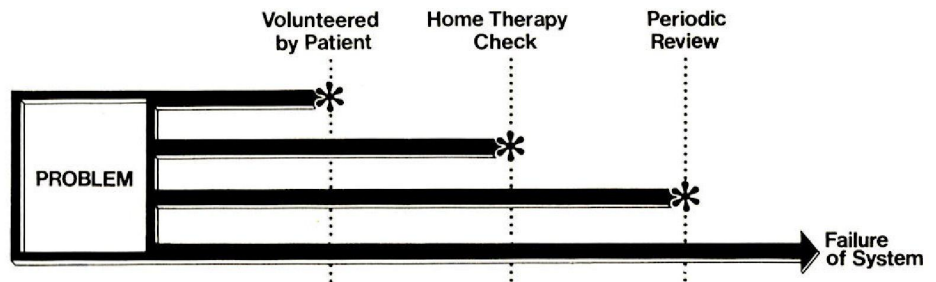
Without efficient structuring of visits, patients may easily be lost to the system. This is detrimental both to the *patients* who, if they are not seen following an acute bleeding episode, may not achieve complete recovery, and to the *Centre staff*, who may lose valuable feedback information which is necessary for assessing the effects of their treatment.

Interim checks

In those Centres to which patients on home therapy must return for their supplies of concentrate, the visits also provide a convenient opportunity for interim checks, including examination for factor antibody and HB_sAg/Ab screening.

Effective organisation

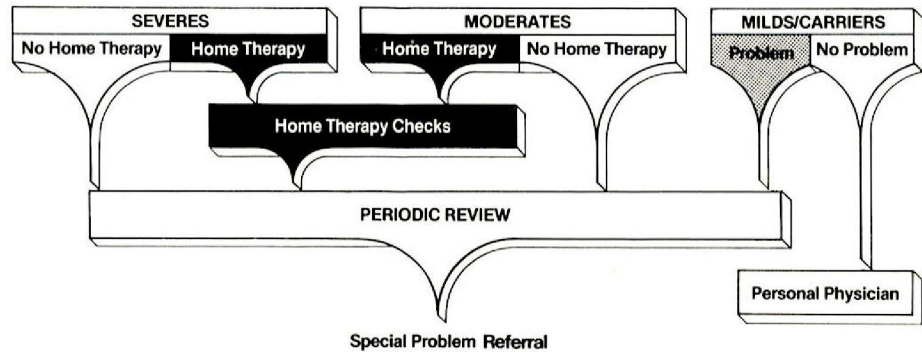
The organisation of the Centre should provide an effective system for recognising and identifying every problem encountered by a severely affected haemophiliac or his family.



If a problem goes undetected this must be regarded as a breakdown in the organisation of the Centre.

Responsibility

The rarity of haemophilia, together with the speed of progress in its management, have occasionally resulted in a dichotomy in the organisation for follow-up. It is suggested that, where a Haemophilia Centre exists, responsibility for follow-up should lie with the staff of that unit, rather than with a personal physician or family doctor.



THE PERIODIC REVIEW CLINIC

The arrangements for the six-monthly and annual review will, of course, vary from Centre to Centre. The recommendations given here are intended as a guide. Whatever the pattern followed in a particular hospital, certain fundamental elements should be common to every follow-up procedure.

Fundamental elements of formal review:

CLINICAL

- General history
- Specific history of bleeding episodes
- General physical examination
- Musculo-skeletal assessment
- Dental assessment
- Social/psychological assessment

INVESTIGATION

- Radiology
- Haematology
- Biochemistry

Recording progress

The method of recording a patient's progress suggested here has been developed in one follow-up clinic. It is used in conjunction with a patient information *base*, recorded on forms designed for the World Federation of Hemophilia. Further details will be found in the booklet on *General Records*.

The record is dependent on an accurate log of every significant bleed and episode of treatment. This log may be kept in a variety of ways, for example in diary form or as a punched card for mechanical sorting. Many home therapy programmes are run on the understanding that further supplies of therapeutic material will only be issued on receipt of an up-to-date log from the patient or his family.

Prior to the follow-up clinic information from the log is sorted and tabulated to give the following essential data, with which recommendations for changes in management may be made.

number of bleeds
frequency of bleeding
sites of bleeding

timing of therapy

administration of therapy

amount of blood product used
type of blood product used

side effects

Examples of tabulation:

Arrow indicates total blood product used in finite period of time (shown at top of form). Ten packs of cryoprecipitate were given — an estimated 700 VIII units.

FOLLOW UP - SUMMARY SINCE LAST ASSESSMENT	
BLOOD PRODUCTS	
Family/Surname BROWN	Hospital No. 614763
Given/First Name(s) ARTHUR	
Record No.	<input type="text"/>
Date last assessment (day/month/year)	<input type="text"/>
Date this assessment (day/month/year)	<input type="text"/>
Blood products used in this interval:	
	VIII/UNITS
Packs blood/red cells	<input type="text"/>
Packs fresh frozen plasma	<input type="text"/>
Packs cryoprecipitate	<input type="text"/>
(i) Estimate Factor Units	<input type="text"/>
Concentrates 1 HEMOPIL	<input type="text"/>
2 LOCAL AHG	<input type="text"/>
3	<input type="text"/>
4	<input type="text"/>
5	<input type="text"/>
6	<input type="text"/>
(ii) Actual Factor Units	<input type="text"/>
TOTAL FACTOR VIII/UNITS	<input type="text"/>
TOTAL ESTIMATED AND ACTUAL	<input type="text"/>

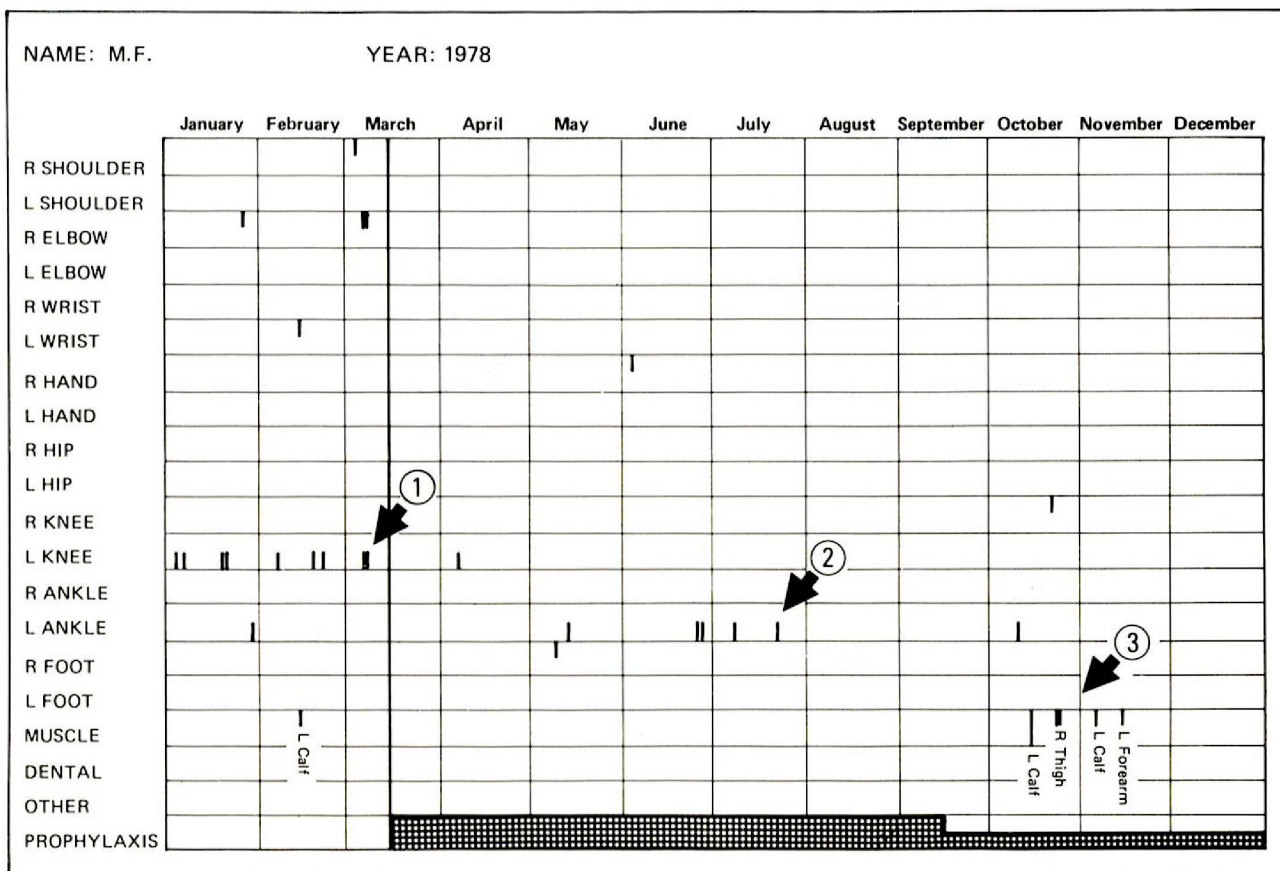
Treatment within			
1 HR	2 HRS	2-12 HRS	>12 HRS
	1	11	2
Total Fresh Bleeding Episodes			14
Treatment Given By:			
Hospital Staff			1
Private Physician			
Relative/Guardian			11
Self			2
Trauma			4
Side Effects			
Mild			-
Severe			-
Analgesia			-
Treatment Satisfactory			13
Treatment Unsatisfactory			1

Upper arrow indicates that 11 injections were given between 2 and 12 hours after onset of bleeding. Why?
Lower arrow indicates that only 2 injections in this series were self-administered. Why?

FOLLOW UP			
Family/Surname BROWN		Hospital No.	
Given/First Name ARTHUR		579603	
WH Record No.			
BLEEDS			
Date last assessment (day/month/year)		11/07/6	
Date this assessment (day/month/year)		14/07/7	
Number of bleeding episodes in this interval			
Joints		Right	Left
shoulder		2	
elbow		1	
wrist			
hip			
knee		2	
ankle		9	1
Muscles			
forearm			
hand			
iliotibial		1	
gluteal			
quadriceps			
hamstrings			1
calf			
Other bleeds, specify: HAEMATURIA X3			
CURRENT ASSESSMENT			
CHANGES SINCE PREVIOUS ASSESSMENT		RECOMMENDATIONS	
1	TARGET R. ANKLE	PLASTIC SPLINT; INCREASE Dose VIT	
2	REC. HAEMATURIA	IVP	
3			
4			
5			
6			

Upper arrow indicates TARGET JOINT – in this case the right ankle.
Lower arrow indicates 3 episodes of haematuria since last follow-up.

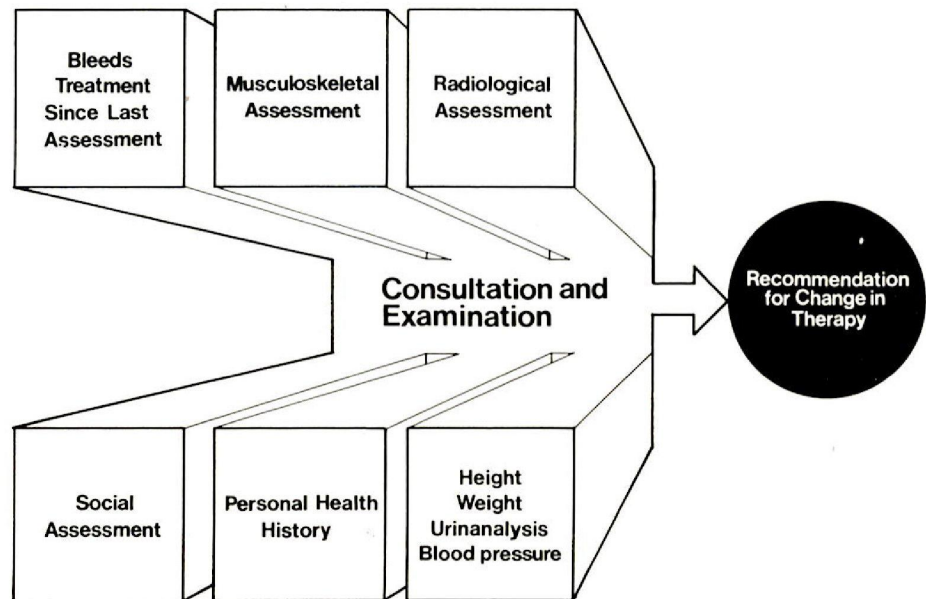
A very useful visual aid to follow-up is the calendar method of recording. Each treatment recorded in the log is marked according to the date and the site of the bleed.



First arrow (March) indicates a run of bleeds in left knee, for which prophylactic factor VIII therapy is prescribed (vertical line). Second arrow (July/August) indicates frequent haemarthroses of left ankle, and subsequent fitting of plastic ankle splint. Third arrow (November) indicates series of muscle injuries — result of start of football season in UK.

Recommendations for changes in therapy

In order to be able to recommend changes in treatment, or to advise on a patient's lifestyle, the Centre doctor must consult several sources of information, including those considered above.



Discussion with patient

This feedback and information from the general physical, musculo-skeletal and radiological profiles should be discussed with the patient, if he is old enough, and with his family, so that the reasons for any recommended changes in treatment are understood. The intelligent patient soon begins to monitor his own progress.

Privacy

However a haemophilia follow-up clinic is arranged, absolute privacy is essential for each family at some stage. Students and paramedical staff should be excluded from at least one consultation with the doctor or with the social worker.

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Scandinavian Journal of Haematology Supplement 31, 37, 1977

10.

GENERAL RECORDS

10

GENERAL RECORDS

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Routine hospital records were designed for short-stay, minimum follow-up patients. As a consequence recording the medical and social features of people with a chronic disorder like haemophilia results in the rapid accumulation of several kilograms of paper, from which it is difficult to extract useful, and often urgently required, information.

Several systems have been devised in an attempt to solve this problem; the introduction of the problem orientated medical record (POMR) by Professor Lawrence Weed provides perhaps the best known example.

L L Weed, 1969, Medical Records, Medical Education and Patient Care. Case Western Reserve University Press.

With the rapid development of microelectronics, computer-based systems may become cheap enough for general application, but they will, of course, still rely on accurate and relevant data input. This in turn relies on a logical system of enquiry.

In the case of haemophilia, enquiry into both past history and present state may best be performed by using special-purpose record sheets. These have two functions:

- they act as checklists for the doctor;
- they eliminate the need for much unnecessary writing.

The special-purpose records presented here are those developed for the World Federation of Hemophilia.

They incorporate the ideas of many doctors working with haemophilia and features of systems developed for individual Centres.

Also shown are examples of other special-purpose sheets kindly provided by their authors.

PART ONE

DIAGNOSIS

The following proformae are used during
diagnostic consultation in the Newcastle
Haemophilia Centre

NORTHERN REGIONAL HAEMOPHILIA SERVICE

Diagnostic Consultation

NAME:		Telephone:	
ADDRESS:			
Referring Hospital/Ward/Consultant			
Study Reference	1-3	A J	
Coagulation Register Number	4-8		
Referred from: University Hospital Group = 1 G.P. = 3			
Regional Hospital = 2 Dentist = 4			
Other = 5	9		
Date of Examination	10-15		
Age (in years and months)			
Date of Birth	16-21		
Sex: Male = 1 Female = 2	22		
Employment			
G.P.	Dentist:		
Reasons for Referral:			
Bruising Yes = 1 No = 2	23		
Purpura Yes = 1 No = 2	24		
Haemorrhage Site(s) Yes = 1 No = 2	25		
Swollen Joint(s) Site(s) Yes = 1 No = 2	26		
Past history bleeding Yes = 1 No = 2	27		
Family history bleeding Yes = 1 No = 2	28		
Other Yes = 1 No = 2	29		
Other illness Yes = 1 No = 2	30		
Diagnoses: Established: (& where made) Provisional:			
HISTORY			
1. Bruising: Excessive = 1 Not Excessive = 2	31		
Always associated with trauma = 1			
Sometimes spontaneous = 2	32		
Superficial bruising: Large (>5 cms. diam.) = 1			
Small (<5 cms. diam.) = 2	33		
Flat = 1 Raised = 2	34		
Deep haematomata: Experienced = 1			
Never = 2	35		
Principle sites of bruising:-			

2. Purpura		Experienced = 1		
Sites affected:		Never = 2	36	<input type="checkbox"/>
3. Lacerations	Experienced	= 1	Never	= 2
	Sutures	= 1	No sutures	= 2
	Prolonged or secondary haemorrhage		= 1	
	No prolonged or secondary haemorrhage		= 2	39
4. Joints (especially knees, ankles, elbows)		Affected = 1		
Sites affected:		Never = 2	40	<input type="checkbox"/>
5. Epistaxes	Excessive	= 1	Never	= 3
	Occasional	= 2		
	If 1 or 2:			41
	Right nostril	= 1	Both	= 3
	Left nostril	= 2	Not known	= 4
			42	<input type="checkbox"/>
6. Gastro-intestinal	Haematemesis	Yes = 1	No = 2	43
	Melaena	Yes = 1	No = 2	44
Associated diagnosis:				
7. Haematuria		Yes = 1	No = 2	45
Associated diagnosis:				
8. Gynae/Obstetric	Applicable	= 1	Not applicable	= 2
	Duration of Periods (days)			46
	Pads/tampons (average 24 hours)			47-48
	Are/were periods considered heavier than normal (when off all therapy)?			49-50
		Yes = 1	No = 2	51
	Pre-menopause	= 1		
	Menopause	= 2		
	Post-menopause	= 3		
	Post-menopausal bleeding=	4		52
	Pregnant	Yes = 1	No = 2	53
	Number of previous pregnancies			54-55
	Post partum haemorrhage:	Yes = 1	No = 2	56
	Sterile	Yes = 1	No = 2	57
	9. Dental extractions		Yes = 1	No = 2
Bleeding following extractions:				
Not excessive		= 1		
Excessive; prolonged > 6 hours		= 2		
Excessive; secondary haemorrhage		= 3		
2 & 3		= 4	59	<input type="checkbox"/>
If 2, 3 or 4 has every extraction been followed by excessive haemorrhage?		Yes = 1 No = 2	60	<input type="checkbox"/>
If 2, 3 or 4 give details of treatment:				

10. Operations Details (age at operation and hospital):	Yes = 1 No = 2	61	<input type="checkbox"/>
Associated undue haemorrhage or wound haematoma	Yes = 1 No = 2	62	<input type="checkbox"/>
11. Transfusions (blood products) Details (include type of product):	Yes = 1 No = 2	63	<input type="checkbox"/>
12. Any other evidence abnormal haemorrhage (neonatal, retroperitoneal, subdural, etc.) Details:	Yes = 1 No = 2	64	<input type="checkbox"/>
13. Does patient take aspirin? Frequently = 1 Occasionally = 2 Never = 3 Has aspirin been taken in past 2 weeks Product(s):-	Yes = 1 No = 2	65 66	<input type="checkbox"/> <input type="checkbox"/>
14. Drugs other than aspirin taken in past 2 weeks: (incl. contraceptive pill)	Yes = 1 No = 2	67	<input type="checkbox"/>
Family history of possible bleeding disorder If yes enter details of affected relatives and where investigated:	Yes = 1 No = 2	68	<input type="checkbox"/>
Names of patient's spouse/children/siblings:			

EXAMINATION	Telangiectasia	Yes = 1 No = 2	69	<input type="checkbox"/>
Specify:	Abnormal Bruising	Yes = 1 No = 2	70	<input type="checkbox"/>
Specify:	Purpura	Yes = 1 No = 2	71	<input type="checkbox"/>
Specify:	Haemarthroses	Yes = 1 No = 2	72	<input type="checkbox"/>
Specify:	Other evidence of bleeding disorder	Yes = 1 No = 2	73	<input type="checkbox"/>
Specify:	Evidence in relative attending consultation	Yes = 1 No = 2	74	<input type="checkbox"/>
Specify:	Evidence other disease	Yes = 1 No = 2	75	<input type="checkbox"/>

Laboratory Results.

•	WBC x10 ⁹ /l
•	RBC x10 ¹² /l
•	Hb g/dl
•	HCT l/l
	MCV fl
•	MCH pg
•	MCHC g/dl

Bleeding Time			secs		FACTOR ASSAY	
W.B.C.T.			secs		I	%
Platelets			/mm ³		II	%
					V	%
TEST	PATIENT	CONTROL			VII	%
R.C.T.	secs	secs			VIII	%
A.D.P.	secs	secs			IX	%
Prothrombin	secs	secs			X	%
Prothrombin Ratio		≡	%		XI	%
P.T.T.K.	secs	secs			XII	%
Thrombin Time	secs	secs			XIII	%
+ Sal. Sulph.	secs	secs			F.D.P.	ug/ml
+ E.A.C.A.	secs	secs			F.D.P. + Reptilase	ug/ml
					E.L.T.	
					INHIB v

Comment:

Diagnosis:

76-77 ☐

Reports to: Referring doctor
Dentist
Family doctor
Other

Signed:

Date:

Positive diagnosis: Entry to Haemophilia Centre Register (date)

PART TWO

PATIENT INFORMATION BASE

The base is completed on admission of a patient to the register of haemophiliacs on confirmation of positive diagnosis.

HAEMOPHILIA CENTRE		CONFIDENTIAL MEDICAL RECORD	
Name:	(Maiden Name):	Record Number:	
Address:		Telephone:	
Name of spouse/children/siblings:			
If family history names of other families affected:			
Date of birth Sex Marital status Religion Nationality Next of kin		<div style="border: 1px solid black; padding: 5px; margin: 0 auto; width: 100px;"> PHOTOGRAPH </div> <div style="border: 1px solid black; padding: 2px; margin: 2px auto; width: 100px;"> Date </div>	
Diagnosis Factor Level Factor Antibody Blood Group Animal AHG in the past? Allergies/sensitivities		Other illness: (1) (2) (3)	
Family Physician Address		Dentist Address	
Telephone		Telephone	
School/employment Address		Ambulance Address	
Telephone		Telephone	
Other hospital/physician involved with patient's treatment Address			
Telephone			

HAEMOPHILIA CENTRE		BASE – DIAGNOSIS CLINICAL AND LABORATORY HISTORY	
NAME			
STUDY REFERENCE CARD REFERENCE RECORD NUMBER	1-2	B	C
	3	1	
	4-9		
DATE OF BIRTH DATE BASE COMPILED	(day/month/year) (day/month/year)	10-15 16-21	
PRIMARY DIAGNOSIS	1 Haemophilia A 2 Haemophilia B 3 von Willebrand's 4 Other (specify)	22	
DEFICIENT FACTOR LEVEL (C/ACTIVITY) %		23-25	
RELATED ANTIGEN LEVEL (RAG/PROTEIN) %		26-28	
BLEEDING TIME (seconds) Method:		29-32	
OTHER LABORATORY EVIDENCE BLEEDING DISORDER (specify) Yes 1 No 2		33	
CLINICAL SEVERITY	1 Severe 2 Moderate 3 Mild	34	
FACTOR ANTIBODY (specify)		35	
BLOOD GROUP	A B AB O Rh Pos Rh Neg	36 37 38 39 40 41	
<div style="border: 1px solid black; padding: 5px; margin: 0 auto; width: 150px;"> (write here) </div>			
Blood grouping problems/irregular antibodies:			

Known exposure to concentrate from (code Yes 1 No 2)	human: voluntary blood human: paid donors animal: porcine animal: bovine	42 43 44 45
Usual administration of blood product in past by: (code Yes 1 No 2)	hospital staff private physician relative or guardian self	46 47 48 49
Usual mode of treatment:	hospital therapy: on demand 1 home therapy: prophylactic 3	50 51
Previous history reactions to blood products (code Yes 1 No 2) Specify:		52
Acute allergic pulmonary oedema in past (code Yes 1 No 2) Specify:		53
STUDY REFERENCE CARD REFERENCE RECORD NUMBER		1-2 3 4-9
DATE OF BIRTH DATE BASE COMPILED (day/month/year) (day/month/year)		10-15 16-21
SUMMARY SEVERE/DISABLING BLEEDING EPISODES		
Intracranial:	Yes 1 No 2	22
Face, eye, oral, neck:	Yes 1 No 2	23
Dental:	Yes 1 No 2	24
Recurrent epistaxes:	Yes 1 No 2	25

Joints	shoulder elbow wrist hand hip knee ankle foot Other	26-27 28-29 30-31 32-33 34-35 36-37 38-39 40-41 42-43
Intramuscular	forearm iliopsoas/retroperitoneal gluteal quadriceps hamstrings calf Other	44-45 46-47 48-49 50-51 52-53 54-55 56-57
Previous history fracture: If 1 site(s)	Yes 1 No 2	58
Previous history pseudotumour:	Yes 1 No 2	59
Haematuria:		60
Haematemesis/melaena:		61
Gynaecological:	Applicable 1 Not applicable 2	62
If applicable —	Problem 1 No problem 2	63
Other severe/disabling bleeds:		64

HAEMOPHILIA CENTRE		GENERAL HEALTH		HAEMOPHILIA CENTRE		SOCIAL HISTORY	
NAME		NAME		NAME		NAME	
STUDY REFERENCE	1-2	STUDY REFERENCE	1-2	STUDY REFERENCE	1-2	STUDY REFERENCE	1-2
CARD REFERENCE	3	CARD REFERENCE	3	CARD REFERENCE	3	CARD REFERENCE	3
RECORD NUMBER	4-9	RECORD NUMBER	4-9	RECORD NUMBER	4-9	RECORD NUMBER	4-9
DATE BASE COMPILED (day/month/year)	10-15	DATE BASE COMPILED (day/month/year)	10-15	DATE COMPILED (day/month/year)	10-15	DATE COMPILED (day/month/year)	10-15
Allergies/sensitivities: Chronic pain	Yes 1 No 2 Persistent 1 Intermittent 2 Absent 3	16		(SPECIFY ALL PROBLEMS)	16		
Usual analgesic:	Problem 1 No problem 2	17		EDUCATION	Problem 1 No problem 2	17	
Major illness other than haemostatic abnormality (ICD Code)	1 2 3 4	18-22		LITERACY	Problem 1 No problem 2	18	
Surgical history (other than dental)	Yes 1 No 2 Year Year Year Year	23-26		QUALIFICATIONS	Yes 1 No 2	19	
Immunisation history: (code Yes 1 No 2)	diphtheria pertussis tetanus poliomyelitis measles smallpox typhoid BCG Other: Yes 1 No 2	27-30		EMPLOYMENT	Preschool 1 Nursery/playschool 2 School 3 Full-time higher education 4 Employed 5 Unemployed 6 Retired 7	20	
Other complaints on systemic enquiry:	Yes 1 No 2	31-34		If 5 does employer know diagnosis ? Yes 1 No 2		21	
DENTAL HEALTH	Regular 1 Irregular 2 Nil 3	35		If 5 is there an employment problem ? Yes 1 No 2		22	
Treatment	Yes 1 Total clearance 2 No 3	36		HOUSING	Problem 1 No 2	23	
Last dental check	Routine 1 Occasional 2 Never 3	37		ACCESS TO TELEPHONE	Problem 1 No 2		
Extractions in past? Details if known	Yes 1 No 2 Unknown 3	38					
Use of local anaesthesia		39					
FAMILY HISTORY BLEEDING DISORDER		40					
(Complete family tree and carrier proforma on separate sheet)		41					
		42					
		43					
		44					
		45					

MOBILITY	Problem 1	No problem 2	24	<input type="checkbox"/>
FAMILY STABILITY	Problem 1	No problem 2	25	<input type="checkbox"/>
FAMILY FINANCE	Problem 1	No problem 2	26	<input type="checkbox"/>
ALLOWANCES/BENEFITS	Problem 1	No problem 2	27	<input type="checkbox"/>
ALCOHOL CONSUMPTION	Problem 1	No problem 2	28	<input type="checkbox"/>
DRUG CONSUMPTION	Problem 1	No problem 2	29	<input type="checkbox"/>
TOBACCO CONSUMPTION	Problem 1	No problem 2	30	<input type="checkbox"/>
ACTIVITIES	Problem 1	No problem 2	31	<input type="checkbox"/>
Sports:				
Hobbies:				

DENTAL HEALTH				
NAME:				
STUDY REFERENCE		1-2	H	F
CARD REFERENCE		3		
RECORD NUMBER		4-9		
DATE (day/month/year)		10-15		
Oral hygiene	Unsatisfactory 1	Satisfactory 2	16	<input type="checkbox"/>
Caries	Present untreated 1	Present restored 2	17	<input type="checkbox"/>
	Absent 3			
Periodontal condition	Unsatisfactory 1	Satisfactory 2	18	<input type="checkbox"/>
Occlusion	Unsatisfactory 1	Satisfactory 2	19	<input type="checkbox"/>
Dentures	Required 1	Present unsatisfactory 2	20	<input type="checkbox"/>
	Present satisfactory 3	Absent 4		

HAEMOPHILIA CENTRE

MUSCULOSKELETAL ASSESSMENT

NAME

RECORD NUMBER

DATE

Musculoskeletal measurement in quiescent state (tick normal, enter measurement abnormal).

Patient RIGHT/LEFT handed.

RIGHT LEFT

	Flex	Ext	Abd	Add	Int/r	Ext/r	Flex	Ext	Abd	Add	Int/r	Ext/r
Normal	180	50	180	50	90	90	180	50	180	50	90	90
Patient												
Muscle Power 0-5												

Shoulder

	Flex	Ext	Pron	Sup	Flex	Ext	Pron	Sup
Normal	150	0	90	90	150	0	90	90
Patient								
Muscle Power 0-5								

Elbow

	Flex	Ext	Radial Dev	Ulnar Dev	Flex	Ext	Radial Dev	Ulnar Dev
Normal	70	70	20	30	70	70	20	30
Patient								
Muscle Power 0-5								

Wrist

	Flex	Ext	Abd	Add	Opp	Flex	Ext	Abd	Opp
Patient									

Thumb

	Flex	Ext	Abd	Add	Flex	Ext	Abd	Add
Patient								

Fingers

	0-5
Grip	

RIGHT LEFT

	Flex	Ext	Abd	Add	Int/r	Ext/r	Flex	Ext	Abd	Add	Int/r	Ext/r
Normal	120	30	50	30	45	45	120	30	50	30	45	45
Patient												
Muscle Power 0-5												

Hip

	Flexion	Extension	Flexion	Extension
Normal	135	0	135	0
Patient				
Muscle Power 0-5				

Knee

	Plant Flex	Dors Flex	Inver	Ever	Plant Flex	Dors Flex	Inver	Ever
Normal	50	20	5	5	20	20	5	5
Patient								
Muscle Power 0-5								

Ankle/Foot

	Flexion	Extension	Flexion	Extension
Patient				

Toes

Deformities (contractures, muscle wasting, subluxed joints etc.) present 1 absent 2 ☐

Gait: abnormal 1 normal 2 ☐

Chronic pain:

Chronic synovitis:

CHANGES SINCE PREVIOUS ASSESSMENT:

PHYSIOTHERAPIST'S COMMENTS:

PHYSICAL EXAMINATION / FOLLOW-UP (Specify all abnormalities)													
NAME:	<table border="1"> <tr> <td>1-2</td> <td>H</td> <td>F</td> </tr> <tr> <td>3</td> <td></td> <td></td> </tr> <tr> <td>4-9</td> <td></td> <td></td> </tr> <tr> <td>10-15</td> <td></td> <td></td> </tr> </table>	1-2	H	F	3			4-9			10-15		
1-2	H	F											
3													
4-9													
10-15													
STUDY REFERENCE CARD REFERENCE RECORD NUMBER DATE (day/month/year)													
AGE NOW	OCCUPATION NOW												
HEIGHT (CMS)	16-18												
WEIGHT (KGS)	19-21												
URINE	22												
	Normal 1												
BLOOD PRESSURE	23-25 26-28												
	SYSTOLIC DIASTOLIC												
STATE OF VEINS	29												
	No problem 2												
LYMPHADENOPATHY	30												
	Yes 1 No 2												
HEPATOMEGALY	31												
	Yes 1 No 2												
SPLENOMEGALY	32												
	Yes 1 No 2												
OTHER ABNORMAL FINDINGS	33												
	Yes 1 No 2												
LAST DENTAL CHECK:	34												
	Problem 1 No Problem 2												

NAME	RECORD NO.
SUMMARY TARGET JOINTS (record relevant dates by affected joints.)	

HAEMOPHILIA CENTRE		RADIOLOGICAL AND LABORATORY RESULTS	
NAME			
STUDY REFERENCE	1-2	B	C
CARD REFERENCE	3	5	
RECORD NUMBER	4-9		
DATE COMPILED (day/month/year)	10-15		
RADIOLOGY			
Routine chest	1 Normal 2 Abnormal 3 Not performed	16	
X-ray (code 1 normal 2 early arthropathy 3 advanced arthropathy 4 not performed)	wrist elbow shoulder hip knee ankle other:	17-18 19-20 21-22 23-24 25-26 27-28 29-30	R L
IVP	1 Normal 2 Abnormal 3 Not performed	31	
LABORATORY			
FBC	1 Normal 2 Abnormal 3 Not performed	32	
Liver function tests	1 Normal 2 Abnormal 3 Not performed	33	
Hb _s Ag	1 Negative 2 Positive 3 Not performed	34	

Hb ₂ Ab	1 Negative 2 Positive 3 Not performed	35	
Coagulation factor ANTIBODY (inhibitor) screen.	1 Negative 2 Equivocal 3 Positive 4 Not performed	36	
Other abnormalities	Yes 1 No 2	37	

PART THREE

PROGRESS

Some suggestions for recording progress are given in Booklet 9 on *Follow-up*.

The following illustrations are of a punched card in use in Newcastle, both by patients on home therapy and by hospital staff.

They are used in conjunction with a calendar, on which the days when the patient is incapacitated are marked.

Physical examination (including musculo-skeletal assessment) forms and radiological and laboratory forms are the same as those illustrated in part two (Patient Information Base).

PART FOUR

HAEMOPHILIA CARRIER

REGISTER

Folding file-card for confidential genetic register.

FAMILY TREE:

1

OBLIGATORY/POSSIBLE CARRIER (VIII/IX)

NAME:

MAIDEN NAME/PREVIOUS NAMES:

ADDRESS:

TELEPHONE:

DATE OF BIRTH:

Day Month Year

RELATIONSHIP TO HAEMOPHILIC:

FAMILY PHYSICIAN:

OBSTETRICIAN/GYNAECOLOGIST:

2

HISTORY ABNORMAL HAEMOSTASIS:

LABORATORY INVESTIGATION:

cp = contraceptive pill + = pregnant: give gestation
RATIO C : RA_g (MEANS) :
RATIO C : RA_g (MEDIANS) :

PROGNOSIS GIVEN:

Fold

Reproductive/potentially reproductive/non-reproductive at time prognosis given.

COMMENTS:

OUTCOME:

[illegible]

PART FIVE

EXAMPLES OF OTHER FORMS

- A Follow-up at Worcester Hemophilia Center, Mass; courtesy of Dr Peter Levine.
- B Bleeding records and follow-up forms in use at Centre 'Air et Soleil', La Queue-lez-Yvelines; courtesy of Dr Jean-Pierre Allain.
- C Example of simple log provided by a commercial company.
- D Running-record type of log designed for computer processing; courtesy of Professor Ilsley Ingram, St Thomas' Hospital, London.
- E Example of running-record of laboratory test results; courtesy of Dr Louis Aledort, Mount Sinai Hospital, New York.
- F Example of Register, including basic information on each patient; courtesy of US National Hemophilia Foundation.
- G Example of simplified, basic instructions to junior doctors/paramedical staff/nursing staff on each patient likely to attend for treatment; courtesy of Dr Peter Levine, Worcester, Mass..
- H Example of patient log of bleeds and therapy; courtesy of the late Dr Katherine Dormandy, Royal Free Hospital, London.
- I Log of bleeds and therapy including details of absence from work/school; courtesy of Dr A E Weiss, Good Samaritan Hospital, Cincinnati.
- J Example of Kardex system adapted for haemophilia record use; courtesy of Dr Stein Evensen, Rikshospitalet, Oslo.
- K Plasma products administration log; courtesy of Dr S L Dietrich, Orthopaedic Hospital, Los Angeles.

A

ANNUAL REPORT — Form A

RECORD (1) 4 Y Y M M D D Date 2 Patient Number 7 Date of Birth 11 16
(DUPLICATE COLS. 2-16 IN ALL RECORDS)

Name 17 Occupation 37 Date begun Self-Therapy 47 Y Y M M D D
Type of Hemophilia: A 1 B 8 Severity of Hemophilia: Mild 10 Moderate 11 Severe 12 VIII or IX Level 13
Mo/Yr. of Dx 14 Associated Illnesses Found

1. Hypertension: Yes 15 No 16 Blood Pressure 17 18 19 20 21 22 23 24 25 26 27 28 29 30
2. Anemia: Yes 31 No 32 Iron Deficiency: Yes 33 No 34
Other Cause: 35 36 37 38 39 40 41 42 43 44 45 46 47 48 49 50

3. Dental Care: None 51 Mild 52 Moderate 53 Severe 54
Extractions performed during past year: None 55 Number 56
4. Surgery performed past year: 57 58 59 60 61 62 63 64 65 66 67 68 69 70 71 72 73 74 75 76 77 78 79 80 81 82 83 84 85 86 87 88 89 90 91 92 93 94 95 96 97 98 99 100

5. Other Illnesses Found: A 101 102 103 104 105 106 107 108 109 110 111 112 113 114 115 116 117 118 119 120

RECORD (2) 3 Y Y M M D D Date 2 Patient Number 7 Date of Birth 11 16
(DUPLICATE COLS. 2-16 IN ALL RECORDS)

Name 17 Occupation 37 Date begun Self-Therapy 47 Y Y M M D D
Type of Hemophilia: A 1 B 8 Severity of Hemophilia: Mild 10 Moderate 11 Severe 12 VIII or IX Level 13
Mo/Yr. of Dx 14 Associated Illnesses Found

1. Hypertension: Yes 15 No 16 Blood Pressure 17 18 19 20 21 22 23 24 25 26 27 28 29 30
2. Anemia: Yes 31 No 32 Iron Deficiency: Yes 33 No 34
Other Cause: 35 36 37 38 39 40 41 42 43 44 45 46 47 48 49 50

3. Dental Care: None 51 Mild 52 Moderate 53 Severe 54
Extractions performed during past year: None 55 Number 56
4. Surgery performed past year: 57 58 59 60 61 62 63 64 65 66 67 68 69 70 71 72 73 74 75 76 77 78 79 80 81 82 83 84 85 86 87 88 89 90 91 92 93 94 95 96 97 98 99 100

5. Other Illnesses Found: A 101 102 103 104 105 106 107 108 109 110 111 112 113 114 115 116 117 118 119 120

LABORATORY DATA

HGB 121 g% Retic 122 % Serum Iron 123 mg% TIBC 124 mg%
HCT 125 % Microcytic Mild 126 Microcytic Moderate 127 Microcytic Severe 128 Macrocytic 129
BLOOD SMEAR: Normochromic 130 Hypochromic Mild 131 Hypochromic Moderate 132 Hypochromic Severe 133

HEPATITIS ASSOC. ANTIGEN: Yes 134 No 135 Titer I/ 136 Titer II/ 137
ASSOC. ANTIBODY: Yes 138 No 139 ALK. PHOS. 140 141 142 143 144 145 146 147 148 149 150

SGPT 151 152 153 154 155 156 157 158 159 160 161 162 163 164 165 166 167 168 169 170

Factor VIII Inhibitor: Yes 171 No 172 Titer 173 174 175 176 177 178 179 180 181 182 183 184 185 186 187 188 189 190

BIURUBIN: Total 191 192 193 194 195 196 197 198 199 200 201 202 203 204 205 206 207 208 209 210

A

ANNUAL REPORT — Form B

RECORD (4) 1 Y Y M M D D Date 2 Patient Number 7 Date of Birth 11 16
(DUPLICATE COLS. 2-16 IN ALL RECORDS)

Patient Name 17 CHRONIC SYNOVITIS: Mild 18 Severe 19 PAIN: Mild 20 Severe 21 CHRONIC SYNOVITIS: Mild 22 Severe 23

R. Ankle 24 L. Ankle 25 R. Knee 26 L. Knee 27 R. Hip 28 L. Hip 29 R. Elbow 30 L. Elbow 31 R. Shoulder 32 L. Shoulder 33

STABILITY OF KNEE IN DEGREES: Varus 34 Valgus 35 Posterior Subluxation 36 Anterior Subluxation 37

RANGE OF MOTION IN DEGREES: Right Knee 38 Left Knee 39 Right Flexion 40 Left Flexion 41 Right Extension 42 Left Extension 43

KNEE: Dorsiflexion 44 Plantar Flexion 45 Flexion 46 Extension 47 Pronation 48 Supination 49

ANKLE: Dorsiflexion 50 Plantar Flexion 51 Flexion 52 Extension 53 Pronation 54 Supination 55

ELBOW: Flexion 56 Extension 57 Pronation 58 Supination 59

SHOULDER: Flexion 60 Abduction 61 Internal Rotation 62 External Rotation 63

HIP: Flexion 64 Abduction 65 Internal Rotation 66 External Rotation 67 Extension 68

LEG LENGTH (inches): 69 70 71 72 73 74 75 76 77 78 79 80 81 82 83 84 85 86 87 88 89 90 91 92 93 94 95 96 97 98 99 100

[illegible]

SCAPULO-HUMÉRALE

- Anté-pulsion
- Rétro-pulsion
- Abduction
- Adduction
- Rotation interne
- Rotation externe

COUDE

- Flexion
- Extension
- Pro-supination

POIGNET


- Flexion dorsale
- Flexion palmaire
- Inclinaison radiale
- Inclinaison cubitale

DOIGTS

Flexion
Extension

Périmètres des Membres Supérieurs

BRAS A

AVANT-BRAS 

[illegible]

B

[illegible]

COXO-FÉMORALE

- Flexion
- Extension
- Abduction
- Adduction
- Rotation interne
- Rotation externe

GENOU

Flexion
Extension

TIBIO-TARSIENNE

Genou étendu	Flexion dorsale
	Flexion plantaire
Genou fléchi	Flexion dorsale
	Flexion plantaire

Périmètres des Membres Inférieurs

CUISSE A

JAMBE à

[illegible]

OBSERVATIONS

PERSONAL
LOGBOOK OF

Month/Year

23

D

E

Bleeding Disorders - Clinic Sheet									
COMMENTS:									
NAME	C B NO.	DATE dd/mm/yy	1-7	8-13	14	15-19	20-24	25-31	32-37
WARD	TIME	ONSET	DATE	TYPE	CONTINUATION	SITE	QTY.	PRODUCT	QTY.
TREATMENT									
DECLARED ACT.									
ASSAYED ACT.									
ROUTE A/D DOSE FREQ. QTY. DRUG CODE									
DRUG APPROVED NAME									

Bleeding Disorders - Clinic Sheet									
COMMENTS:									
NAME	C B NO.	DATE dd/mm/yy	1-7	8-13	14	15-19	20-24	25-31	32-37
WARD	TIME	ONSET	DATE	TYPE	CONTINUATION	SITE	QTY.	PRODUCT	QTY.
TREATMENT									
DECLARED ACT.									
ASSAYED ACT.									
ROUTE A/D DOSE FREQ. QTY. DRUG CODE									
DRUG APPROVED NAME									

Bleeding Disorders - Clinic Sheet									
COMMENTS:									
NAME	C B NO.	DATE dd/mm/yy	1-7	8-13	14	15-19	20-24	25-31	32-37
WARD	TIME	ONSET	DATE	TYPE	CONTINUATION	SITE	QTY.	PRODUCT	QTY.
TREATMENT									
DECLARED ACT.									
ASSAYED ACT.									
ROUTE A/D DOSE FREQ. QTY. DRUG CODE									
DRUG APPROVED NAME									

NEW YORK REGIONAL HEMOPHILIA CARE SYSTEM
HEPATITIS TESTS RECORD

REC. ID.	DATE																ACCOUNT NO.															
1 2 3	4	5	6	7	8	9	10	11	12	13	14	15	16	17	18	19	20	21	22	23	24	25	26	27	28	29	30	31	32	33		
H 2 0																																

ANTI HBS																ANTI H																ANTI BC																ANTI AG																ANTI AB																SGPT																																																																															
HA																BS																C																C																C																C																																																																															
24																25																26																27																28																29																30																31																32																33															

F

PATIENT REGISTRY FORM			
Instructions: Please print information requested in the space provided or circle the most appropriate number. (Circle only one number for each question.)			
BASIC DATA:			
1. Patient's name:	7. Physician's telephone number:		
Last Name	Area Code	Number	
First Name	Middle Initial	8. Physician's specialty	
2. Patient's address:		1. Hematologist	
Number Street		2. Pediatrician	
City	State	3. Internist	
Zip		4. General Practitioner	
3. Patient's telephone number:		5. Other (Specify)	
Area Code	Number	9. Hospital or Treatment Center usually visited for care:	
4. If patient is under 18, name of parent or guardian:		Name of Hospital/Treatment Center	
Last Name		10. Address of Treatment Center:	
First Name	Middle Initial	Number Street	City State Zip
5. Physician with major treatment responsibility:		11. Is patient/family affiliated with the National Hemophilia Foundation?	
Last Name		1 Yes	
First Name	Middle Initial	2 No (If no, skip to # 13)	
6. Physician's address:		12. If affiliated, what is the name of the Chapter?	
Number Street		Name of NHF Chapter	
City	State	13. Patient's social security number:	
Zip		Social Security Number	

PERSONAL DATA:	
14. Patient's sex:	20. Does patient have a family history of hemophilia?
1 Male	1 Yes
2 Female	2 No
	3 Unknown
15. Patient's year of birth:	21. Is patient presently in school?
Year of birth	1 Yes
	2 No
16. Patient's siblings (Ever born)	22. Highest year of school or grade patient has attended (Circle one):
Number of sisters	Preschool or has never attended
Number of brothers	0
	Elementary through high school
17. Patient's marital status:	1 2 3 4 5 6 7 8 9 10 11 12
1 Single	College
2 Married	13 14 15 16 17 18 or more
3 Widowed	
4 Divorced	
18. Patient's children (Ever born)	23. If patient is out of school, is he presently employed?
Number of daughters	1 Yes
Number of sons	2 No (If no, skip to # 26)
	24. If yes, give patient's job title:
19. Patient's race/ethnic background:	Job Title
1 White	
2 Negro or black	
3 Spanish surname or descent	
4 Indian (Amer.)	
5 Chinese	
6 Japanese	
7 Filipino	
8 Hawaiian	
9 Other (Specify)	
	25. Yearly family income:
	1 Under \$5,000
	2 \$5,000 - 9,999
	3 \$10,000 - 14,999
	4 \$15,000 - 19,999
	5 \$20,000 - 24,999
	6 \$25,000 Or over

MEDICAL DATA: Please verify answers given in this section with physician		INSURANCE DATA:	
26. Patient's blood group:	33. Is primary mode of treatment?	38. Is patient covered by health (Medical) insurance?	42. CONT'D.
1 A	1 In response to bleeding	1 Yes	Medicare
2 B	2 To prevent bleeding (prophylaxis)	2 No	1 Yes
3 AB		3 Unknown	2 No
4 0	34. Is patient in a home therapy (self-infusion) program?		Medicaid
5 Unknown	1 Yes		1 Yes
	2 No		2 No
27. Patient's Rh:	35. Blood product most often used during the past 12 months (Circle one only)		Welfare
1 Positive	1 Wet-frozen cryoprecipitate (Cryo)		1 Yes
2 Negative	2 Factor VIII dry concentrate		2 No
3 Unknown	3 Factor IX dry concentrate		1 Yes
	4 Plasma		2 No
28. Patient's factor deficiency (Type of hemophilia):	5 Whole blood		State-sponsored hemophilia program
1 Factor VIII (Classic Type A)	6 Other (Specify)		1 Yes
2 Factor IX (Christmas Type B)	7 Unknown		2 No
3 Factor XI			Other agency or program
4 Von Willebrand's disease	36. Give approximate numbers for the past twelve months for each of the following: (If none write zero. If unknown leave blank.):		1 Yes (Specify)
5 Other (Specify)	Number of infusions patient received		2 No
6 Unknown	Number of outpatients visits (Emergency rooms, clinics, etc.)		
	Number of times hospitalized for hemophilia bleeding		43. If patient received assistance from one or more of the above during the past 12 months, was it for
29. Severity of deficiency:	Total number of days hospitalized for the above		Blood and blood products
1 Mild	Number of times hospitalized problems related to hemophilia other than bleeding		1 Yes
2 Moderate	Total number of days hospitalized for the above		2 No
3 Severe	Number of days missed from school or work for hemophilia-related reasons		3 Unknown
4 Unknown			Medical Care
30. Factor level of deficiency:			1 Yes
1 0-1%			2 No
2 1-2%			3 Unknown
3 2-3%			Hospitalization
4 3-5%			1 Yes
5 5-30%			2 No
6 30-50%			3 Unknown
7 Over 50%			41. Has patient received assistance in past 12 months from some government program or agency?
8 Unknown			1 Yes
31. Has patient been tested for an inhibitor (antibody)?			2 No
1 Yes			3 Unknown
2 No			Other
3 Unknown			1 Yes (Specify)
32. If yes, did latest test show presence of an inhibitor (antibody)?			2 No
1 Yes			3 Unknown
2 No			44. Please estimate the total amount family spent (out of pocket) for patient's hemophilia care in past 12 months:
3 Unknown			Amount \$

F

MEDICAL DATA: Please verify answers given in this section with physician		INSURANCE DATA:	
26. Patient's blood group:	33. Is primary mode of treatment?	38. Is patient covered by health (Medical) insurance?	42. CONT'D.
1 A	1 In response to bleeding	1 Yes	Medicare
2 B	2 To prevent bleeding (prophylaxis)	2 No	1 Yes
3 AB		3 Unknown	2 No
4 0	34. Is patient in a home therapy (self-infusion) program?		Medicaid
5 Unknown	1 Yes		1 Yes
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4 Von Willebrand's disease	36. Give approximate numbers for the past twelve months for each of the following: (If none write zero. If unknown leave blank.):		1 Yes (Specify)
5 Other (Specify)	Number of infusions patient received		2 No
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29. Severity of deficiency:	Number of times hospitalized for hemophilia bleeding		43. If patient received assistance from one or more of the above during the past 12 months, was it for
1 Mild	Total number of days hospitalized for the above		Blood and blood products
2 Moderate	Number of times hospitalized problems related to hemophilia other than bleeding		1 Yes
3 Severe	Total number of days hospitalized for the above		2 No
4 Unknown	Number of days missed from school or work for hemophilia-related reasons		3 Unknown
30. Factor level of deficiency:			Medical Care
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2 1-2%			2 No
3 2-3%			3 Unknown
4 3-5%			Hospitalization
5 5-30%			1 Yes
6 30-50%			2 No
7 Over 50%			3 Unknown
8 Unknown			41. Has patient received assistance in past 12 months from some government program or agency?
31. Has patient been tested for an inhibitor (antibody)?			1 Yes
1 Yes			2 No
2 No			3 Unknown
3 Unknown			Other
32. If yes, did latest test show presence of an inhibitor (antibody)?			1 Yes (Specify)
1 Yes			2 No
2 No			3 Unknown
3 Unknown			44. Please estimate the total amount family spent (out of pocket) for patient's hemophilia care in past 12 months:
			Amount \$

F

<p>45. This form was completed by (Circle one)</p> <ul style="list-style-type: none"> 1 Patient 2 Parent 3 Physician 4 Chapter staff 5 Treatment Center Staff 6 Other (Specify) 	<p>50. for ID card, give name of person to contact in emergency:</p> <p>Last Name</p> <p>First Name Middle initial</p>
<p>46. Has information been verified by</p> <p>Physician</p> <ul style="list-style-type: none"> 1 Yes 2 No <p>Treatment Center</p> <ul style="list-style-type: none"> 1 Yes 2 No 	<p>51. Telephone number of above contact person:</p> <p>Area Code Number</p>
<p>47. Has a Registry form like this previously been filled out for this patient?</p> <ul style="list-style-type: none"> 1 Yes 2 No (If no or unknown, skip to # 49) 3 Unknown 	<p>Thank you for giving me the opportunity to register and to provide confidential data for national statistics on hemophilia.</p>
<p>48. If yes, what is patient's hemophilia Registry number?</p>	<p>Signature of patient (or parent, if under 18)</p>
<p>49. Please send me a Hemophilia Registry Identification card.</p> <ul style="list-style-type: none"> 1 Yes 2 No <p>If no, skip # 50 and # 51</p>	<p>Date _____</p> <p>month/day/year</p>

HEMOPHILIA

Date of Completion of this Card _____

Name _____ Date of Birth _____ N.E.M.C.H. # _____

Address _____ Phone _____

Diagnosis _____ Severity _____ Inhibitor _____

Blood Type _____ Irregular Antibodies _____

Attending Physician _____ M.D. Phones _____

_____ M.D. Phones _____

_____ M.D. Phones _____

Hematology Fellow on Call _____ M.D. Phones _____

(obtain via N.E.M.C.H. telephone operator)

Standing Orders (See back of this card) (Note that doses grow with the patient; standing orders for growing boys may be obsolete)

Infusion Dosage

Minor Episodes _____

Major Episodes _____

Medications _____

Reactions to Transfusions _____

Comments _____

N.B. There are three identical copies of this card

1. Attending physician's office

2. Hemophilia Club

3. Emergency Room

ANY ALTERATIONS MUST BE MADE TO ALL 3 COPIES,
AND DATED!

G

TREATMENT										
HOSPITAL NUMBER SURNAME:				FIRST NAMES:				HAEMOPHILIA CENTRE DIAGNOSIS:		
DATE	TIME	PLACE (HOME/OP/IP)	BY WHOM	REASON FOR INJECTION	DRUGS	INJECTION MATERIAL X bags/bottles	UNITS/ BLOOD GROUP	VOLUME (mls)	SERIAL/ BATCH NOS.	EFFECTS/COMMENTS

H

Hemophilia Home Treatment Program
Good Samaritan Hospital
Cincinnati, Ohio 45220

TRANSFUSION RECORD

Name _____

Transfusion Date & Time	Site of Bleeding	Time After Symptoms First Noticed	Concentrate Type & Amount	Duration of Symptoms After Transfusion	Absence from School/Work	Reaction (Describe)	Problems/Comments

Navn:		Fødselsnr.:																										
Hemofili <input type="checkbox"/>		Adresse																										
Faktornivå/år:																												
Alvorlig <input type="checkbox"/>	Inhibitor/år:	Sivilstand	Tlf.																									
Moderat <input type="checkbox"/>	Hjemmetransfusjon/år:	Kommune	Fylke																									
Mild <input type="checkbox"/>	Id.kort/ar	Trygdekonto																										
Foresatte, navn/adr.:		Tlf.																										
Mors navn/adr.:		Tlf.																										
Fars navn/adr.:		Tlf.																										
Arb.plass mor/far:		Tlf.																										
Første henvendelse dato/fra:																												
Arvelighetsskjema/dato:		Fullmakt:																										
Sykehus:	Adr.:	Tlf.:																										
Sykehusleger:																												
Privatlege:	Adr.:	Tlf.:																										
Fysioterapeut:	Adr.:	Tlf.:																										
Tannlege:	Adr.:	Tlf.:																										
Medlem av bløderforeningen:																												
År	Hjemme- besøk	Observasjons- opphold	<table border="1"> <tr> <th colspan="2">Bosatt internat</th> <th colspan="2">Bosatt vernet hybel</th> <th colspan="3">Behandlingsleir</th> <th rowspan="2">Andre tilbud</th> </tr> <tr> <th>Fra</th> <th>Til</th> <th>Fra</th> <th>Til</th> <th>Tilbud</th> <th>Ja</th> <th>Nei</th> <th>Delt.</th> </tr> <tr> <td> </td> <td> </td> <td> </td> <td> </td> <td> </td> <td> </td> <td> </td> <td> </td> <td> </td> </tr> </table>	Bosatt internat		Bosatt vernet hybel		Behandlingsleir			Andre tilbud	Fra	Til	Fra	Til	Tilbud	Ja	Nei	Delt.									
Bosatt internat		Bosatt vernet hybel		Behandlingsleir			Andre tilbud																					
Fra	Til	Fra	Til	Tilbud	Ja	Nei		Delt.																				

Ny pas.	Død i år	Diagnose				Måned for neste kontakt												
		A	B	VW	Andre	1	2	3	4	5	6	7	8	9	10	11	12	N.år
Navn:		Født:																

KARDEX BESTEM PÅ SKALAEN HVOR RESPEKTIVE SYNLIGE RUBRIKKER SKAL BEGYNNE.
SKRIV SÅ NÆR PERFORERINGEN SOM MULIG. RIV DERETTER AV TALONGEN.

KARDEX SYSTEM ½ - OSLO 1
KONTORORGANISASJON
PRINSENGT. 6 - SENTRALBORD 41 98 40

J

NAME	# OF BOTTLES DISPENSED												OPD #		DATE	
	DATE		DATE		DATE		DATE		DATE		DATE		DATE		DATE	
AREA OF BLEEDING	R	L	R	L	R	L	R	L	R	L	R	L	R	L	R	L
ELBOW																
KNEE																
SHOULDER																
HIP																
WRIST																
ANKLE																
HAND																
FOOT																
RETROPERITONEAL																
BEHIND KNEE																
FOREARM																
CALF																
OTHER AREAS (SPECIFY)																
HEAD																
NECK																
THROAT																
NOSE																
MOUTH																
FACE																
INTESTINAL																
URINARY																
OTHER (SPECIFY)																
HOW LONG HAVE YOU BEEN BLEEDING?																
NAME OF CONCENTRATE USED																
# OF BOTTLES USED																
LOT #																
# OF UNITS PER BOTTLE																
MEDICATIONS, PROBLEMS OR COMMENTS IF ANY																
SIGNATURE																

OPD NO. _____ B/D _____

NAME _____ HEMO TYPE _____ LEVEL _____ BLOOD TYPE _____

K

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11. GUIDELINES FOR ORGANISATION

11

GUIDELINES FOR ORGANISATION

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“ A Haemophilia Centre is an instrument to provide comprehensive therapy and long-term planning. The Centre should command certain minimum facilities, including the ability to perform the necessary laboratory tests, to provide in-patient care, to supply the necessary products for replacement therapy and to keep appropriate records. ”

This is the definition given to the concept of a Haemophilia Centre in 1976 by an expert group of physicians concerned with haemophilia care, meeting in Luxembourg.

In order to stand ready to provide care of this kind, the group specified the minimum staff required by a Centre:

- the Centre Director;
- an Orthopaedic Surgeon;
- a Nurse;
- a Social Worker;
- a Physiotherapist;
- Laboratory Personnel.

It was considered that the skills which these people offer must be permanently available. Experience in the therapy of haemophilia, especially on the part of the Director and the Orthopaedic Surgeon, was considered by the group to be essential.

The very nature of haemophilia demands a commitment on the part of the Centre staff, not to the haemophiliac alone, but to his whole family.



The haemophiliac is part of a family. His problems must always be treated in this context.

The haemophilic patient and his family have five basic needs:

- accurate diagnosis;
- immediate access to quality blood products and experienced service;
- skilled counselling;
- fast and effective communication;
- 24-hour cover.

These needs can only be fulfilled by a Haemophilia Centre staffed by a team of medical and paramedical personnel experienced in haemophilia management. This by no means precludes the services of the personal physician or family doctor, nor does it mean that haemophiliacs should not receive treatment in smaller local hospitals.

It should be emphasised, however, that any such arrangements should be formalised, so that there is always a link with the nearest Haemophilia Centre.

It is important for this link with the Centre to be maintained after the implementation of home therapy. It is true that treatment at home reinforces the independence of the family and serves the haemophiliac's needs in the community far better than a hospital programme, but it does *not* guard the patient against complications like antibody development, advancing haemophilic arthropathy, and the appearance of side effects to intensive transfusion therapy.

HAEMOPHILIA CENTRE

Functions

The extent of the services offered by a Haemophilia Centre will obviously be related to the number of severely affected haemophiliacs requiring regular treatment. The staff, however, should always be prepared to treat bleeds *immediately and at any time*.

There is a conflict between the necessity for fast treatment near the patient's home and the need for the provision of the costly back-up facilities required for major surgery and the management of complications. In the United Kingdom a solution to this problem has been attempted by grouping Centres by geographical administrative region, each major region — with a population of three to six million — being served by a Reference Centre.

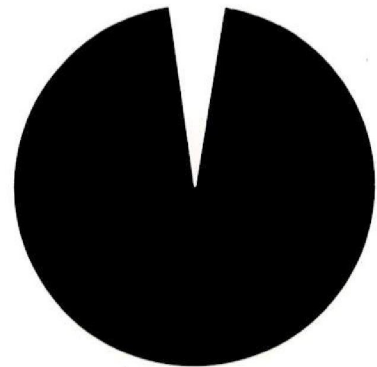
The functions of the smaller Centres vary according to the interests of the staff and the facilities available.



MAJOR FUNCTIONS OF THE REFERENCE CENTRE:

COMPREHENSIVE LABORATORY SERVICE FOR DIAGNOSIS AND MONITORING
ROUND-THE-CLOCK COVER
HOME THERAPY SUPERVISION
FAST OUT-PATIENT CONSULTATIVE SERVICE
IN-PATIENT FACILITIES FOR ALL AGES
COMPREHENSIVE FOLLOW-UP FACILITIES
ORTHOPAEDIC AND GENERAL SURGICAL SERVICES
DENTAL SERVICES INCLUDING CONSERVATION
CARRIER DETECTION SERVICE/GENETIC COUNSELLING
GYNAECOLOGICAL/OBSTETRIC SERVICES INCLUDING FAMILY PLANNING
SOCIAL SERVICES
PSYCHOLOGICAL/PSYCHIATRIC SERVICES
PHYSIOTHERAPY: PHYSICAL MEDICINE/HYDROTHERAPY/REHABILITATION THERAPY
THERAPEUTIC MONITORING/SUPPLY OF QUALITY BLOOD PRODUCTS
RESEARCH
TEACHING

The Centre must not only be regarded as providing a reliable emergency service, but it must also be organised to serve the haemophiliac and his family in the community.



The white section of this pie-chart represents the proportion of his life which a haemophiliac is likely to spend under medical care. The remainder of his time is spent in the community, where he needs constant support from the Centre staff.

Links A Haemophilia Centre cannot work in isolation. Its two most obvious links must be with a *Department of Haematology* and with a *reliable source of blood products*.

Links should also be forged with the appropriate *financial authority* responsible for funding the costly treatment required in a comprehensive care programme, and with the *educational, career guidance and employment authorities*.

In many areas members of *voluntary organisations*, including the *Haemophilia Society*, play an active role in the maintenance of an efficient Centre.

Staffing The level of staffing depends on the number of severely affected patients involved. It is suggested that the minimum staffing requirement (part-time) of an Associated Haemophilia Centre should be:

- a Haematologist;
- an Adult Physician;
- a Paediatrician;
- a Dental Surgeon.

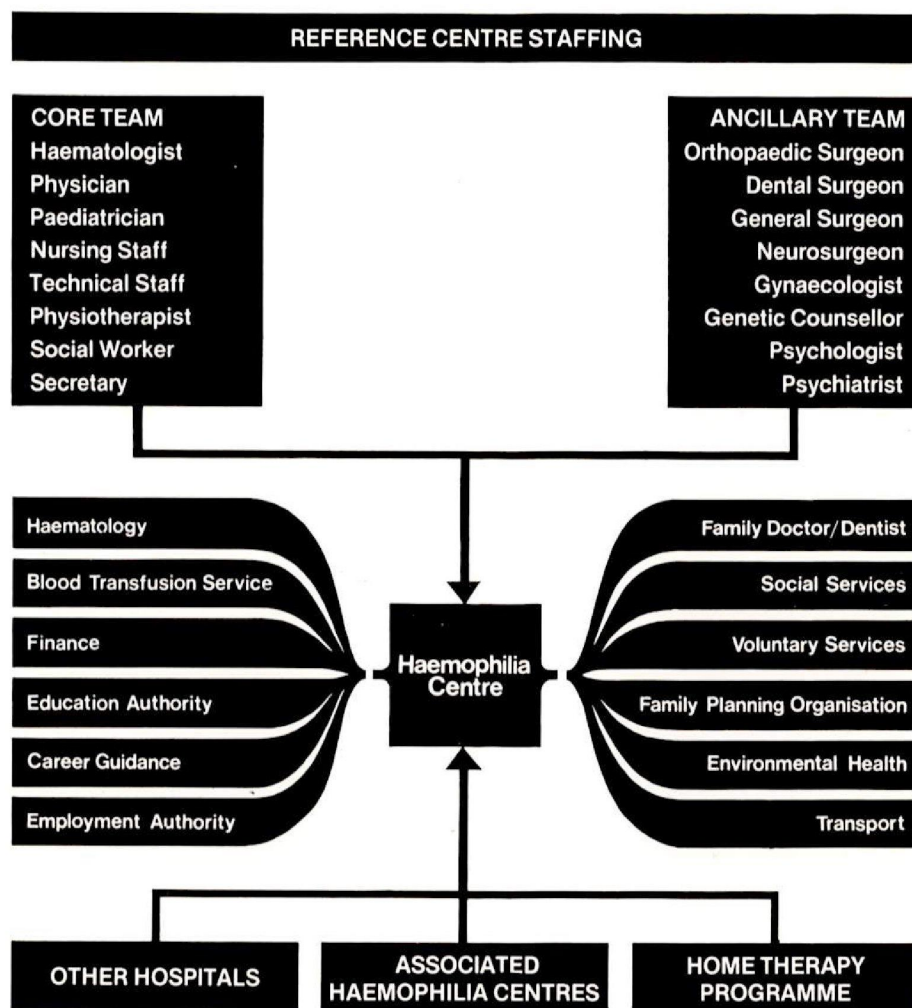
In addition to these, a Reference Centre will require the following staff with an interest and experience in haemostatic failure:

- a General Surgeon;
- a Neurosurgeon;
- a Gynaecologist;
- a Psychologist;
- a Psychiatrist;
- Secretarial Staff.

The Director should be a clinical haematologist or internist or paediatrician with the necessary experience in haemophilia management.

The make-up of the *core* and *ancillary* or back-up teams will vary from Centre to Centre.

In general the core team is composed of those people likely to be involved in the day-to-day management of patients, and the ancillary team comprises those specialists who, aware of the special problems of haemophilia, may be called in to deal with specific problems.



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U K Department of Health and Social Security

Health Circular H C (1976) 4