Objectives

1 to determine on a national base,

- the number of patients affected with these severe haemorrhagic disorders and their geographical repartition
- the most significant circumstances of treatment (severe bleedings, surgery)
- the treatments applied (type of concentrate, amounts of units, CED) and their modalities (proportion of ambulatory treatment, prophylaxis...)

a surveillance system able to immediately investigate any suspicion of transmission of emerging new agents related to the treatments.

Criteria for inclusion

- defect (<30%) in FVIII or FIX
- severe defect (<10%) in FII, V, VII, X, XI, XIII
- severe defect (<0.1 g/l) in fibrinogen
- severe defect in VWF defined as bleeding time > 15 min
  and/or FVIII:C <20%
  and/or VWF:RCO <10%

Participating centres

Organisation

- Periodically (about once a year), centres send information on each patient (anonymously) recording on enrolment his (her) main characteristics and antecedents and on each survey the main events occurred since the last record. The patients are informed of the study but are not requested to sign a special form.
- Data are collected and monitored by a coordinating* centre attached to a public health organisation (InVS).
- A web site (http://www.francecoag.org) is already operational to provide information on the protocol, to load special forms, answer to questions (FAQ)… with a private part restricted to active participants. Direct transmission of patient’s forms via the web site is under development.
- A steering committee follows up the project: clinicians, coordinators, committed health organisations, scientific experts, representatives of patients’ associations.
- The study started in January 2003 as replacement of a previous project dedicated only to haemophilia (Suivi National des Hémophiles) (Calvez et al **)

Results

Number

Type of pathology*

Age at inclusion

0 - 4 years
5 - 10 years
11 - 20 years
> 20 years

3120 patients

- haemophilia A (n=2040)
- haemophilia B (n=422)
- WWD (n=230)
- other (n=86)

- FVIII defect (n=23)
- FIX defect (n=14)
- Anti-fibrinogenemia (n=14)
- FX defect (n=6)

- 0 - 4 years
- 5 - 10 years
- 11 - 20 years
- > 20 years

Comments

Beside its epidemiological interest this project will help to promote transversal research on a well characterized and controlled cohort of rare bleeding disorders (Pups study as example, cf poster by H Chambost)

* On 2784 forms analyzed at that time