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#### AIM OF THE STUDY

Behçet disease (BD) is exceptionally observed before the age of 16 years and raises diagnosis problems because there is no specific biological marker. Sets of clinical criteria have been proposed for adult patients. However, those criteria are not specific for children in whom the disease is often uncompleted or atypical.

The aim of this study is to set-up an international cohort of patients selected on homogenous criteria established by a committee of experts, in order to help further epidemiological studies. The cohort is aimed also to support genetic studies.

#### SCIENTIFIC COMMITTEE

### AIM OF THE STUDY

### SCIENTIFIC COMMITTEE

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#### THE DATABASE

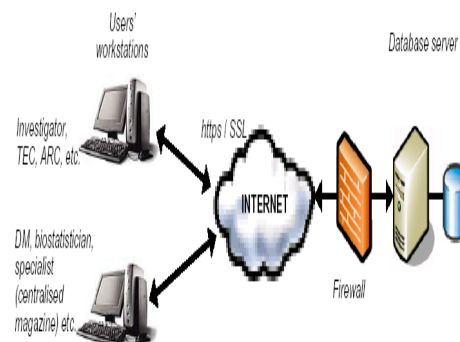
### THE DATA BASE

The data base consists on an electronic case report form (e-CFR).

The CleanWEB™ connector supplies the user interface, allows to access data, and enables him to carry out must data entry task.

The CleanWEB™ connector communicates with the server as well as permits to save datas in the server database for « Synchronisation ».

The investigators only have access to patient data included in their centre according to the access rights which are granted.



▪ SCHEDULE

Centers from all over Europe, specializing in pediatric BD are expected to collaborate under the auspices of the PReS (Pediatric Rheumatology European Society) and the ISBD (international society for Behçet's disease), to document their patients into a single database.

▪ The scientific committee, during the last PReS meeting, has established a list of calling signs (minimal requirement), on consensus basis, in order to define the criteria for entering the study. The participants have reviewed all items of patient charts in details. Long-term documentation is requested.

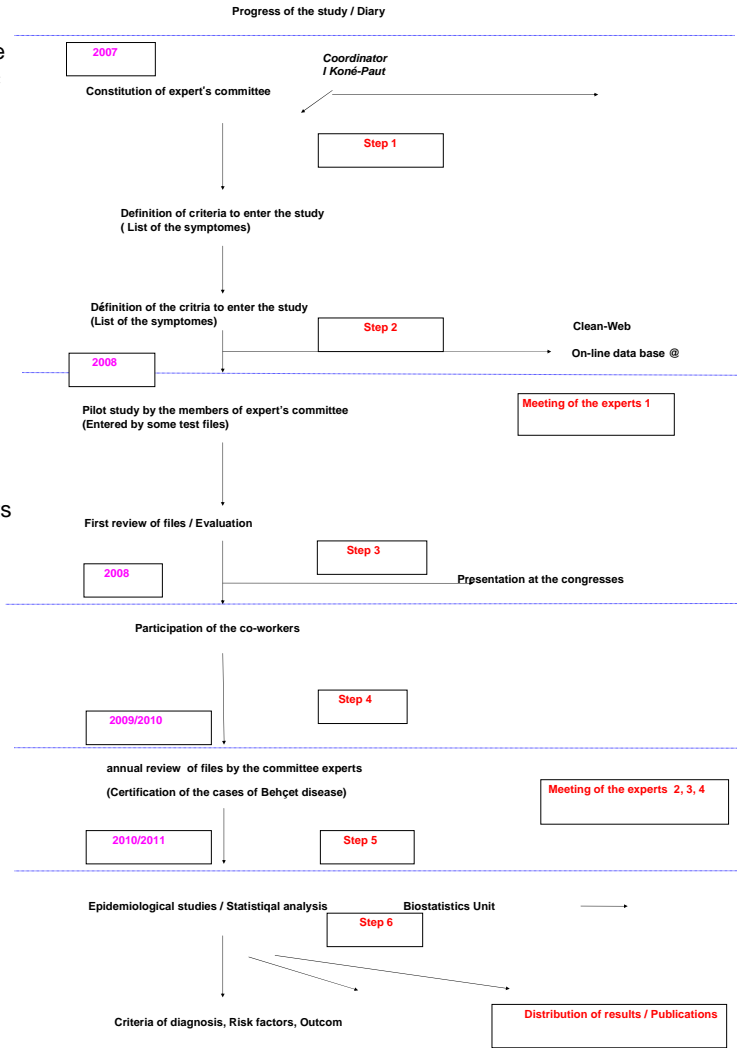
▪ Data will be annually examined by the international scientific committee of experts and the system will recall you each year, automatically, to update your patient data.

▪ The study has received an ethical committee agreement. Informed consent will be obtained for anonymous collection of data and DNAs (according to the legislation of each participating country).

▪ *Criteria of judgment* Patients charts will be reviewed each year by the expert committee in order to classify them as definite BD (consensus), probable BD (majority of votes), or not BD.

*Statistics* Univariate and multivariate analyses will compared each groups of patients and will allow the calculation by symptoms of risks for developing BD.

SCHEDULE



▪ CRITERIA

HOME PAGE OF THE CRITERIA TO ENTER THE DATABASE

**CleanWeb - PEDBD - PUPH-CS Isabelle KONÉ-PAUT - Coordinateur**

Patients Suivi Données Gestion Aide

Nouveau patient A faire

Indicateurs: A compléter

**Criteria to enter the database - Patient non-inclus**

- Inclusion:**
  - Criteria to enter the datab:
  - Demographic data
  - First visit and retrospective
  - Follow-up (year 1)
  - Follow-up (year 2)
  - Follow-up (year 3)
  - Follow-up (year 4)
  - Genetic data and associate
  - Last visit and experts comm
- Hors planning:**
  - Intermediate visit
- Criteria to enter the database - Patient non-inclus:**
  - Informed consent signed  yes  no
  - Planned follow-up for at least 4 years  yes  no
  - Symptoms:** Please list all the symptoms the patient has ever experienced. (He/She should have at least an oral aphthosis plus one of the other following signs)
    - Buccal aphthosis (recurrent or relapsing)  yes  no
    - Genital aphthosis  yes  no
    - Erythema nodosum  yes  no
    - Skin ulceration  yes  no
    - Necrotic folliculitis, pustular or acneiform lesion  yes  no
    - Pathergy phenomenon  yes  no
    - Uveitis  yes  no
    - Retinal vasculitis  yes  no
    - Venous thrombosis  yes  no
    - Arterial thrombosis  yes  no
    - Arterial aneurism  yes  no
    - Familial history of Behçet disease  yes  no  unknown
  - Number of symptoms: 0

**CleanWeb - PEDBD - PUPH-CS Isabelle KONÉ-PAUT - Coordinateur**

Patients Suivi Données Gestion Aide

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Indicateurs: A compléter

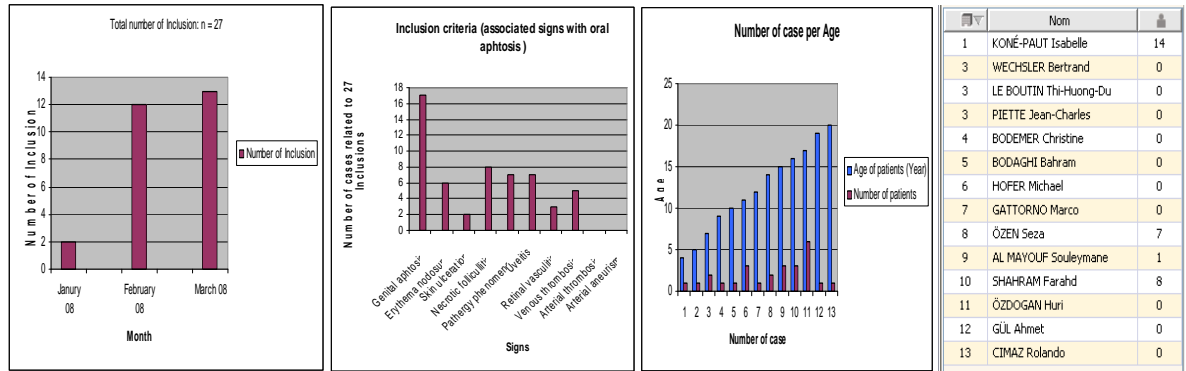
**Demographic data - Patient non-inclus**

- Inclusion:**
  - Criteria to enter the datab:
  - Demographic data
  - First visit and retrospective
  - Follow-up (year 1)
  - Follow-up (year 2)
  - Follow-up (year 3)
  - Follow-up (year 4)
  - Genetic data and associate
  - Last visit and experts comm
- Hors planning:**
  - Intermediate visit
- Demographic data - Patient non-inclus:**
  - Initial Last Name (Initial Nom)
  - Initial First name (Initiale Prénom)
  - Birth date (MM / YYYY)  /
  - Sex  Male  Female
  - Native country of the father (Enter the first letters)
  - Native country of the mother (Enter the first letters)
  - Consanguinity  No  Yes
  - Familial case  No  Yes
  - Other inflammatory disorder of the patient or of his relatives  no inflammatory disorder  personal case  Familial case

▪ **RECRUITMENT SUCCESS**

**LAUNCH OF THE STUDY & RECRUITMENT SUCCESS**

- A total of 36 patients have been included in the data base.
- Thanks and congratulations for all participating investigators.
- Preliminary data :



▪ **HIGHLIGHTS**

**HIGHLIGHTS**

- We wish to motivate all the investigators to include the maximum of patients.
- For the investigators who did not begin yet, we hope to see their participation soon. Mr. Sylla, the clinical research associate, is available for any type of support.
- The launch of the study study was recently presented in the congress of Rome by Pr. Isabelle Kone-Paut. We had the pleasure to see a new investigator joining us following this presentation.
- In the approach of the 13<sup>th</sup> International Conference and 5<sup>th</sup> Patient's Convention for Behçet's Disease organised by the International Society For Behçet's Disease (ISBD) which will held in May 24-27, 2008 in Pörschach (Austria), we wish all investigators to include their eligible patients as much as possible. We plan to present all updated data in this congress.

▪ **REMARK**

**REMARK**

In this newsletter, we aim to outline statutory aspects appropriate for every country concerning clinical research :

- Investigators have to be sure to respect clinical research rules of their country
- Investigators have to obtain : Informed consent signed for the duration of the study (at least 4 years).
- The patient must be a new patient or a patient followed for suspected BD for less than 3 years.
- A patient blood sample as well as his two relatives (parents) must be collected and sent to the genetics molecular laboratory of the hospital of La Timone in Marseille (France)

▪ **DISCUSSION**

**DISCUSSION**

Inclusions have not yet started for many centers due to the delay for obtaining agreement from local ethical committees.  
We hope that these centers are going to begin to include as fast as possible!

▪ **PERSPECTIVES**

**PERSPECTIVES**

This research designed to define tools for early diagnosis of BD in children is mandatory for further epidemiological studies on this topic. In the future, the prospective registration of new cases will permit better understanding of its natural history, its outcome (morbidity, mortality) and its prognosis thanks to long term follow-up and to identify risk factors. Its specific focus on pediatric cases will favor the identification of genetic factors that will be useful for earlier diagnosis of this severe and complex disease.