

## **PED-BD: An international cohort study for pediatric Behçet's disease**

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### **Rationale :**

The Behçet disease (BD) is exceptionally observed before the age of 16 years and raises diagnosis problems because there is no specific biologic marker. Sets of clinical criteria have been proposed for adult patients only. They lack of specificity in children where the disease is often uncompleted or atypical.

### **Aim of the study :**

This research aims to set-up an international cohort of patients selected on homogenous criteria established by a committee of experts<sup>(\*)</sup>, in order to help further epidemiological studies. The cohort is aimed also to support genetic studies

### **Patients and methods :**

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 (An electronic case reported form) that is available online. The scientific committee, during the last PReS meeting, has been 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 are periodically 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) from patients and parents (trios). *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.

### **Agenda :**

The study is ongoing from January 2008.(see the attached newsletters)

### **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.

### **Please join us!! :**

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**(\*) Here the list of inclusion criteria :**

### **General**

- Behçet's disease is suspected
- Disease onset is before 16 years
- The patient is a new patient or the first visit was < 3years
- Follow-up is planned for at least 4 years
- Informed consent has been signed (if applicable in your contry)

### **Clinical signs**

The patient should have experienced at least

- Oral aphthosis (recurrent or relapsing at least 3 times/year)

**plus at least one of the other following signs :**

- Familial history of the Behçet disease
- Genital aphtosis
- Erythema nodosum
- Skin ulceration
- Necrotic folliculitis, pustular or acneiform lesion
- Pathergy phenomenon
- Uveitis
- Retinal vasculitis
- Venous thrombosis
- Arterial thrombosis
- Arterial aneurism