

# RaDiCo-ACOSTILL - Adult and pediatric cohort with Still's disease (RaDiCo-ACOSTILL)

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## General

### Identification

Detailed name	Adult and pediatric cohort with Still's disease (RaDiCo-ACOSTILL)
Sign or acronym	RaDiCo-ACOSTILL
CNIL registration number, number and date of CPP agreement, AFSSAPS (French Health Products Safety Agency) authorisation	N° CCTIRS 16-088bis / N° CPP 14128 ND / N° MESR DC-2015-2479 / MR-001

### General Aspects

Medical area	Internal medicine Pediatrics Rare diseases Rheumatology
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Study in connection with Covid-19	No
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Pathology (details)	Adult Still's disease (AOSD) and systemic onset juvenile idiopathic arthritis (SoJIA) represent two rare multifactorial diseases associated with systemic inflammation. These two forms, AOSD and SoJIA, are considered two facets of the same syndrome, combining four cardinal symptoms [high fever > 39°C, arthralgia or arthritis, skin rash, leukocyte formula with more than 80% neutrophil polymorphonuclear cells]; lymphadenopathy and splenomegaly may also be present; there is significant biological inflammatory syndrome with elevated C-reactive protein, serum ferritin with a dramatic decrease in glycosylated fraction. The incidence of the disease is low, around 0.1 / 100,000 for adults and 0.6 / 100,000 for children. Its prevalence is approximately 1 to 3/100,000 and 3/100,000 for children, so there are approximately 500 to 1500 adults and 450 children affected in France. It is subdivided into pediatric and adult forms according to the age of onset before or after
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16 years. The prognosis of the disease is functional and vital. Macrophage activation syndrome (MAS) is frequently associated, either at the onset of the disease, at the initiation of treatment, or concomitant with viral reactivation. The evolution over time has mainly been studied in children and is variable: regression, episodic progression with regression over time, and chronic joint evolution. In adults, these three evolutionary modes can also be observed. However, differences seem to exist between AOSD and SoJIA.

## Scientific investigator(s) (Contact)

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## Collaborations

## Funding

Funding status	Public
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## Details

The RaDiCRaDiCo-ACOSTILL cohort initially received funding from the state managed by the National Research Agency (ANR) as part of the "Investissements d'Avenir" cohorts program (PIA).

## Governance of the database

Sponsor(s) or organisation(s) responsible

National Institute of Health and Medical Research (Inserm)

Organisation status

Public

Presence of scientific or steering committees

Yes

Labelling and database evaluation

Security audit certification of the database. Data management and continuous quality control of data.

## Additional contact

## Main features

## Type of database

Type of database

Morbidity registers

Study databases (details)

Cohort study

Database recruitment is carried out by an intermediary

A selection of health institutions and services

Database recruitment is made on the basis of:

Another treatment or procedure

Database recruitment is carried out as part of an interventional study

No

Additional information regarding sample selection.

All pediatric and adult patients already diagnosed and followed (prevalent patients) or newly diagnosed (incident patients) in one of the French Reference Centers for Rare Diseases or Rare Disease Competence Centers will be invited to participate in the study. In order to document the improvement in patient management, morbidity, and mortality through the implementation of the Still's Disease National Diagnostic and Care Guidelines (PNDS Still), deceased patients may be included in the cohort. The objective is to recruit a minimum of 200 adult patients and 300 pediatric patients to ensure that the study has sufficient

statistical power.

## Database objective

### Main objective

The main objective is to describe the natural history of the disease in adult and pediatric populations.

### Inclusion criteria

The inclusion criteria for the RaDiCo-ACOSTILL cohort are as follows:

- Patients aged over 16 years (age >16 years) meeting the Yamaguchi diagnostic criteria or Fautrel criteria.
- Patients aged 16 years or younger (age ≤16 years) fulfilling the 2001 criteria for systemic onset juvenile idiopathic arthritis according to the ILAR classification.
- Having signed consent to participate in the cohort and for the collection of clinical and biological data; in accordance with regulations, for deceased minors or adults under legal protection, non-opposition from legal representatives will be sought.
- Affiliated with the social security system.

The exclusion criteria are:

- Other causes of recurrent infectious fever (such as tuberculosis, toxoplasmosis, deep abscesses, viral infections, sepsis) or tumor-related fever (such as lymphomas).
- Other defined inflammatory rheumatic diseases such as rheumatoid arthritis, psoriatic arthritis, spondyloarthropathies.
- Autoimmune inflammatory diseases (systemic lupus erythematosus), granulomatosis (sarcoidosis, Blau syndrome), vasculitis (Behçet's disease, polyarteritis nodosa), polymyositis, and dermatomyositis.
- Well-defined autoinflammatory syndromes with unambiguous mutations, such as familial Mediterranean fever, cryopyrinopathies, TRAPS, mevalonate kinase deficiency.
- Known genetic macrophage activation syndromes.
- Patients unable to understand the information leaflet and sign the informed consent form.
- Patients not affiliated with the social security system.

## Population type

### Age

Newborns (birth to 28 days)  
Infant (28 days to 2 years)  
Early childhood (2 to 5 years)  
Childhood (6 to 13 years)

Adolescence (13 to 18 years)  
Adulthood (19 to 24 years)  
Adulthood (25 to 44 years)  
Adulthood (45 to 64 years)  
Elderly (65 to 79 years)  
Great age (80 years and more)

Population covered	Sick population
Pathology	M05-M14 - Inflammatory polyarthropathies
Gender	Male Woman
Geography area	National
<b>Data collection</b>	
<b>Dates</b>	
Date of first collection (YYYY or MM/YYYY)	2017
Date of last collection (YYYY or MM/YYYY)	2027
<b>Size of the database</b>	
Size of the database (number of individuals)	< 500 individuals
Details of the number of individuals	422
<b>Data</b>	
Database activity	Current data collection
Type of data collected	Clinical data Declarative data Paraclinical data Biological data
Clinical data (detail)	Direct physical measures Medical registration
Details of collected clinical data	The main variables collected are: demographics, family and medical history, socio-economic data, clinical and biological data, clinical signs, symptoms, specific disease treatments, and quality of life self-assessment questionnaires.
Declarative data (detail)	Paper self-questionnaire

## Internet self-questionnaire

Details of collected declarative data  
HAQ/CHAQ child and parent / SF 36 - SF10 (adult - child) / Psychological impact (Hamilton, adult) / Impact on work productivity and activity (WPAI, adult) / Perceived impact (PASS MCII, adult)

Paraclinical data (detail)  
Imaging data (standard radiographs of painful joints when performed)

Biological data (detail)  
Biochemical, hematological, and immunological data

Presence of a biobank  
Yes

Contents of biobank  
Whole blood  
Plasma  
DNA

Details of biobank content  
This study includes a collection of biological samples conducted as part of research for future studies (DNA, RNA, plasma, peripheral blood mononuclear cells).

Health parameters studied  
Health event/morbidity  
Health event/mortality  
Health care consumption and services  
Quality of life/health perception  
Others

Care consumption (detail)  
Hospitalization  
Medical/paramedical consultation  
Medicines consumption

## Procedures

Data collection method  
eCRF in secure web access, secure cloud and HADS hosting

Classifications used  
Drug Dictionary (DCIs)

Quality procedure(s) used  
Data Management Plan and Data Validation Plan.  
Continuous data management (automatic control rules and query system)

Participant monitoring  
Yes

Monitoring procedures  
Monitoring by convocation of the participant  
Monitoring by contact with the referring doctor

Links to administrative sources  
No

## Promotion and access

## Promotion

## Access

Presence of document that lists variables and coding procedures Yes

Terms of data access (charter for data provision, format of data, availability delay)

Requests for access to RaDiCo-ACOSTILL data (aggregated or individual) will be reviewed by the Scientific Committee following the submission of a summary of a Specific Research Project, as defined in the Access Charter. Requests should be sent to the following address: [acostill@radico.fr](mailto:acostill@radico.fr)

Access to aggregated data

Access on specific project only

Access to individual data

Access on specific project only