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
Study in connection with Covid-19	No
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

	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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Unit	UMR S 1136
Organization	National Institute of Health and Medical Research (Inserm)
Collaborations	
Funding	
Funding status	Public

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 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

Data collection

Dates

Date of first collection (YYYY or MM/YYYY)	2017
Date of last collection (YYYY or MM/YYYY)	2027

Size of the database

Size of the database (number of individuals)	< 500 individuals
Details of the number of individuals	422

Data

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
Access to aggregated data	Access on specific project only
Access to individual data	Access on specific project only