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

Responsable(s) :Georgin-Lavialle Sophie , UMRS 938
Fautrel Bruno, UMR S 1136

Date de modification : 03/12/2024 | Version : 1 | ID : 74149

Général

Identification

Nom détaillé	Adult and pediatric cohort with Still's disease (RaDiCo-ACOSTILL)
Sigle ou acronyme	RaDiCo-ACOSTILL
Numéro d'enregistrement (ID-RCB ou EUDRACT, CNIL, CPP, etc.)	N° CCTIRS 16-088bis / N° CPP 14128 ND / N° MESR DC-2015-2479 / MR-001

Thématiques générales

Domaine médical	Internal medicine Pediatrics Rare diseases Rheumatology
Etude en lien avec la Covid-19	No
Pathologie, précisions	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.

Responsable(s) scientifique(s)

Nom du responsable	Georgin-Lavialle
Prénom	Sophie
Adresse	Hôpital TENON, 4 rue de la Chine, 75020 PARIS.
Téléphone	0033(0) 1 56 01 60 77
Email	sophie.georgin-lavialle@aphp.fr
Laboratoire	UMRS 938
Organisme	National Institute of Health and Medical Research (Inserm)
Nom du responsable	Fautrel
Prénom	Bruno
Adresse	Groupe Hospitalier Universitaire Pitié-Salpêtrière 83 boulevard de l'Hôpital, 75651 Paris cedex 13, France
Téléphone	+33(0) 1 421 77801
Email	bruno.fautrel@aphp.fr
Laboratoire	UMR S 1136
Organisme	National Institute of Health and Medical Research (Inserm)

Collaborations

Financements

Financements	Public
Précisions	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).
Gouvernance de la base de données	
Organisation(s) responsable(s) ou promoteur	National Institute of Health and Medical Research (Inserm)
Statut de l'organisation	Secteur Public
Existence de comités scientifique ou de pilotage	Yes
Labellisations et évaluations de la base de données	Security audit certification of the database. Data management and continuous quality control of data.
Contact(s) supplémentaire(s)	
Caractéristiques	
Type de base de données	
Type de base de données	Morbidity registers
Base de données issues d'enquêtes, précisions	Cohort study
Origine du recrutement des participants	A selection of health institutions and services
Critère de sélection des participants	Another treatment or procedure
Le recrutement dans la base de données s'effectue dans le cadre d'une étude interventionnelle	No
Informations complémentaires concernant la constitution de l'échantillon	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.

Objectif de la base de données	
Objectif principal	The main objective is to describe the natural history of the disease in adult and pediatric populations.
Critères d'inclusion	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.
Type de population	
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 concernée	Sick population
Pathologie	M05-M14 - Inflammatory polyarthropathies
Sexe	Male Woman
Champ géographique	National
Collecte	
Dates	
Année du premier recueil	2017
Année du dernier recueil	2027
Taille de la base de données	
Taille de la base de données (en nombre d'individus)	< 500 individuals
Détail du nombre d'individus	422
Données	
Activité de la base	Current data collection
Type de données recueillies	Clinical data Declarative data Paraclinical data Biological data
Données cliniques, précisions	Direct physical measures Medical registration
Détail des données cliniques recueillies	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.
Données déclaratives, précisions	Paper self-questionnaire Internet self-questionnaire
Détail des données déclaratives	HAQ/CHAQ child and parent / SF 36 - SF10 (adult -

recueillies	child) / Psychological impact (Hamilton, adult) / Impact on work productivity and activity (WPAI, adult) / Perceived impact (PASS MCII, adult)
Données paracliniques, précisions	Imaging data (standard radiographs of painful joints when performed)
Données biologiques, précisions	Biochemical, hematological, and immunological data
Existence d'une biothèque	Yes
Contenu de la biothèque	Whole blood Plasma DNA
Détail des éléments conservés	This study includes a collection of biological samples conducted as part of research for future studies (DNA, RNA, plasma, peripheral blood mononuclear cells).
Paramètres de santé étudiés	Health event/morbidity Health event/mortality Health care consumption and services Quality of life/health perception Others
Consommation de soins, précisions	Hospitalization Medical/paramedical consultation Medicines consumption
Modalités	
Mode de recueil des données	eCRF in secure web access, secure cloud and HADS hosting
Nomenclatures employées	Drug Dictionary (DCIs)
Procédures qualité utilisées	Data Management Plan and Data Validation Plan. Continuous data management (automatic control rules and query system)
Suivi des participants	Yes
Modalités de suivi des participants	Monitoring by convocation of the participant Monitoring by contact with the referring doctor
Appariement avec des sources administratives	No
Valorisation et accès	
Valorisation et accès	

Accès	
Existence d'un document qui répertorie les variables et les modalités de codage	Yes
Charte d'accès aux données (convention de mise à disposition, format de données et délais de mise à disposition)	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
Accès aux données agrégées	Access on specific project only
Accès aux données individuelles	Access on specific project only