

# RaDiCo-PID - Idiopathic Interstitial Pneumonia: From Infancy to Elderly

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## Général

### Identification

Nom détaillé Idiopathic Interstitial Pneumonia: From Infancy to Elderly

Sigle ou acronyme RaDiCo-PID

Numéro d'enregistrement (ID-RCB ou EUDRACT, CNIL, CPP, etc.) CCTIRS n° 16.050Bis

### Thématiques générales

Domaine médical Otolaryngology or ENT  
Pediatrics  
Pneumology  
Rare diseases

Pathologie, précisions Idiopathic Interstitial Pneumonia: Idiopathic Interstitial Pneumonia (IIP), known in French as ? Pneumopathies Interstitielles Diffuses (PID)? and referred in the current protocol as IPP/PID, encompass a group of diffuse infiltrative lung diseases of unknown origin that affect the lung architecture and are characterized by a progressive and often irreversible remodeling of the lung. Clinical expression includes mainly dyspnea, restriction on pulmonary function testing, impaired haematosis and radiologic diffuse lung infiltration. In most situations, these diffuse lung disorders are chronic, with high morbidity and mortality due to the lack of curative therapy.

Déterminants de santé Climate  
Genetic  
Healthcare system and access to health care services  
Lifestyle and behavior  
Medicine  
Occupation  
Pollution  
Social and psychosocial factors

Responsable(s) scientifique(s)

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Organisme	Institut National de la Santé et de la Recherche Médicale / French National Institute for Health and Medical Research (Inserm)

## Collaborations

Participation à des projets, des réseaux, des consortiums Yes

Précisions Filière de Santé Maladies Rares pulmonaires RespiFil / European Reference Network ERN-LUNG

## Financements

Financements Public

Précisions Funded by the French « Investissements d'Avenir » cohorts programme, Grant « ANR » 10-COHO-0003.

## Gouvernance de la base de données

Organisation(s) responsable(s) ou promoteur Institut National de la Santé et de la Recherche Médicale / French National Institute for 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

## Contact(s) supplémentaire(s)

## Caractéristiques

### Type de base de données

Type de base de données

Morbidity registers

Informations complémentaires concernant la constitution de l'échantillon

The goal of the IIP/PID cohort is to include prevalent and incident IIP/PID cases diagnosed in paediatric patients and adult patients. For the prevalent cases and the retrospective nature of the data, a diagnosis validation will be required.

Paediatric patient population

Paediatric IIP/PID patients include all patients with diffuse parenchymal diseases linked to rare conditions others than immune deficiencies, proliferative disorders, metabolic disorders, and drug toxicity.

Since the first description of the RespiRare IIP/PID paediatric cases, almost 400 patients (prevalent cases) have been included in the database. Around 60-80 new IIP/PID cases (incident cases) are currently reported per year. However, this number is underestimated, and will most likely increase with the identification of more adapted diagnostic criteria.

Adult patient population

For IIP/PID patients with IPF (approximately two thirds of the IIP/PID adult patients): considering the relatively large number of patients mainly aged and with a very poor prognosis, only prospective data will be collected to maximize the longitudinal collection of data and allow a detailed and accurate description of disease evolution in this population.

For IIP/PID patients without IPF but with diffuse parenchymal diseases linked to rare conditions others than immune deficiencies, proliferative disorders, metabolic disorders, and drug toxicity (approximately one third of the IIP/PID adult patients): IIP/PID, incident cases and prevalent cases (with retrospective data) will be included in the cohort.

About 2000 adult IIP/PID patients are expected to be recruited during this study.

### Objectif de la base de données

Objectif principal

Primary Objective

The main objective is to describe the phenotypic features of the paediatric and adult patients with IIP/PID, at diagnosis and during the follow-up. These data will be critical for the description of the natural history of the various forms of IIP/PID.

### Secondary Objectives

The secondary objectives are to:

- ? Identify gene factors involved in disease initiation and progression,
- ? Investigate the extent to which environmental and co-morbidity factors may influence disease severity and outcome
- ? Identify and validate biomarkers for disease diagnosis and progression

### Exploratory objectives

- ? Production of improved strategies for patient recruitment and enrolment into clinical trials
- ? Development of novel strategy for patient follow-up
- ? Development of novel diagnostic approaches
- ? Evaluation of effect on natural history of disease, and tolerance of therapy, in a large population in real life
- ? Development of novel therapeutic approaches

### Information Technology Objectives

- ? Develop and diffuse an electronic tool of data collection from various sources linked to a database integrating a system of management and follow-up of data-management allowing collection of data for IIP/PID paediatric and adult patients.
- ? Include data generated by patients and, where relevant, their parents and/or carers.

### Critères d'inclusion

- ? Patient with a diagnosis of IIP/PID  
IIP/PID diagnosis is established on presenting history, clinical, radiological and functional and if available pathological findings. Inclusion criteria include:
  - ? Clinical criteria: chronic respiratory insufficiency manifestations including dyspnea/tachypnea, cough, and cyanosis during exercise or at rest
  - ? Radiological criteria: characteristic chest High-Resolution Computed Tomography (HRCT) abnormalities including widespread ground glass or alveolar attenuation, reticulation often associated with traction bronchiectasis, and honeycombing
  - ? Functional criteria: pulmonary function test abnormalities reflecting a restrictive pattern and including: loss of lung volume, vital capacity (VC), total lung capacity (TLC); reduction in the diffusion capacity of the lung for carbon monoxide (DLCO), gas exchange abnormalities, and altered ventilatory response to exercise
  - ? Patients (parents/guardians for paediatric/patients) having given an informed

consent to participate in the protocol  
? Patients affiliated to the ?Regime National  
d'Assurance Maladie?

#### Non-inclusion Criteria

? Patients with diffuse parenchymal lung diseases  
caused by drug toxicity, immunodeficiency,  
proliferative disorders including histiocytosis, and  
metabolic disorders  
? Patients (parents/guardians for paediatric patient)  
not able to approve/understand the protocol

### 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 J84 - Other interstitial pulmonary diseases

Sexe  
Male  
Woman

Champ géographique National

Détail du champ géographique Complete national coverage through the network of  
rare pulmonary disease reference and competence  
centers

### Collecte

#### Dates

Année du premier recueil 2017

Année du dernier recueil 2021 minimum

### Taille de la base de données

Taille de la base de données (en  
nombre d'individus) [1000-10 000[ individuals

Détail du nombre d'individus 2700

## Données

Activité de la base

Current data collection

Type de données recueillies

Clinical data  
Declarative data  
Paraclinical data  
Biological data  
Administrative data

Données cliniques, précisions

Direct physical measures  
Medical registration

Détail des données cliniques recueillies

This is a national multi-centric non interventional study, built on a model of observational longitudinal study, descriptive, retrospective and prospective allowing to collect homogenous clinical, biological, treatment, environmental, and quality of life data from paediatric and adult patients with idiopathic interstitial pneumonia associated to biological data collection: from blood (DNA samples, serum), biopsies and broncho-alveolar liquid (BAL) analyses.

Données déclaratives, précisions

Paper self-questionnaire  
Internet self-questionnaire  
Face to face interview

Détail des données déclaratives recueillies

SF36 or SF10 + St George's Hospital

Données biologiques, précisions

Record of biological results (hematology, biochemical, immunity, serology); Record of results for: bacteriology, virology, parasitology, mycology, bronchoscopy, bronchoalveolar lavage examination; Record of lung function tests, arterial blood gas and spirometry (If available sleep gas exchange and polysomnography); If available/performed, records of results for lung tissue examination, lung biopsy (surgical, transbronchial), lung explant; If available/performed, records of other organ function evaluation (including digestive and cardiac examinations)

Existence d'une biothèque

Yes

Contenu de la biothèque

Serum  
Fluids (saliva, urine, amniotic fluid, ?)  
Tissues  
DNA  
Others

Détail des éléments conservés

plus broncho alveolar liquids

Paramètres de santé étudiés	Health event/morbidity Health event/mortality Quality of life/health perception Others
<b>Modalités</b>	
Mode de recueil des données	eCRF using REDCap; Cloud based, secure by design, web accessible platform. Certified Health Data Hosting resource
Nomenclatures employées	HPO, ICD10, Snomed CT, Orpha Codes and ORDO, Drug dictionary (DCIs)
Procédures qualité utilisées	Continuous data management; Data Management Plan and Data Validation Plan. Native controls 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 Monitoring by crossing with a medical-administrative database
Appariement avec des sources administratives	No
<b>Valorisation et accès</b>	
<b>Valorisation et accès</b>	
<b>Accès</b>	
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)	Access requests to RaDiCo-PID data (rough / structured), biocollections or to analytic reports will be examined by the scientific committee following submission of a Specific Research Project (SRP) synopsis, as defined in the Resource Access Charter. Must be sent to pid@radico.fr
Accès aux données agrégées	Access on specific project only
Accès aux données individuelles	Access on specific project only