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

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General

Identification

Detailed name	Idiopathic Interstitial Pneumonia: From Infancy to Elderly
Sign or acronym	RaDiCo-PID
CNIL registration number, number and date of CPP agreement, AFSSAPS (French Health Products Safety Agency) authorisation	CCTIRS n° 16.050Bis

General Aspects

Medical area	Otolaryngology or ENT Pediatrics Pneumology Rare diseases
Pathology (details)	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.
Health determinants	Climate Genetic Healthcare system and access to health care services Lifestyle and behavior Medicine Occupation Pollution Social and psychosocial factors

Scientific investigator(s) (Contact)	
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Organization	Institut National de la Santé et de la Recherche Médicale / French National Institute for Health and Medical Research (Inserm)
Collaborations	
Participation in projects, networks and consortia	Yes
Details	Filière de Santé Maladies Rares pulmonaires RespiFil / European Reference Network ERN-LUNG
Funding	
Funding status	Public
Details	Funded by the French « Investissements d'Avenir » cohorts programme, Grant « ANR » 10-COHO-0003.
Governance of the database	
Sponsor(s) or organisation(s) responsible	Institut National de la Santé et de la Recherche Médicale / French National Institute for 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

Additional contact

Main features

Type of database

Type of database	Morbidity registers
Additional information regarding sample selection.	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 Pediatric 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.

Database objective

Main objective	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.
Inclusion criteria	? 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

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	J84 - Other interstitial pulmonary diseases
Gender	Male Woman
Geography area	National
Detail of the geography area	Complete national coverage through the network of rare pulmonary disease reference and ciompetence centers

Data collection

Dates

Date of first collection (YYYY or MM/YYYY)	2017
Date of last collection (YYYY or MM/YYYY)	2021 minimum

Size of the database

Size of the database (number of individuals)	[1000-10 000[individuals
Details of the number of individuals	2700

Data

Database activity	Current data collection
Type of data collected	Clinical data Declarative data Paraclinical data Biological data Administrative data
Clinical data (detail)	Direct physical measures Medical registration
Details of collected clinical data	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.
Declarative data (detail)	Paper self-questionnaire Internet self-questionnaire Face to face interview
Details of collected declarative data	SF36 or SF10 + St George's Hospital
Biological data (detail)	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)
Presence of a biobank	Yes
Contents of biobank	Serum Fluids (saliva, urine, amniotic fluid, ?)

	Tissues DNA Others
Details of biobank content	plus broncho alveolar liquids
Health parameters studied	Health event/morbidity Health event/mortality Quality of life/health perception Others
Procedures	
Data collection method	eCRF using REDCap; Cloud based, secure by design, web accessible platform. Certified Health Data Hosting resource
Classifications used	HPO, ICD10, Snomed CT, Orpha Codes and ORDO, Drug dictionary (DCIs)
Quality procedure(s) used	Continuous data management; Data Management Plan and Data Validation Plan. Native controls and Query system
Participant monitoring	Yes
Monitoring procedures	Monitoring by convocation of the participant Monitoring by contact with the referring doctor Monitoring by crossing with a medical-administrative database
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)	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
Access to aggregated data	Access on specific project only
Access to individual data	Access on specific project only