Site ID I__I__I	Patient ID I__I__I	Patient Initials I__I__I__I

CHARACTERISTICS AND MANAGEMENT OF PATIENTS WITH CTD RELATED ILD NATIONAL - MULTICENTRIC DATABASE

CASE REPORT FORM

Participating Physician	……
Site Name	……
Site ID	I__I__I
Patient Initials	I__I__I__I
Patient ID	I__I__I

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	INCLUSION CRITERIAS	YES	NO
1	Patients aged 18 years old or older	☐	☐
2	Patients evaluated as CTD-ILD between January 2018 and December 2019.	☐	☐

ALL ANSWERS HAVE TO BE YES

	EXCLUSION CRITERIAS	YES	NO
1	Patients under 18 years.	☐	☐

ANSWER HAS TO BE NO

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Data Collection Date	I__I__I / I__I__I / I__I__I__I__I

DEMOGRAPHICS

Date of Birth	I__I__I / I__I__I / I__I__I__I__I
Gender	☐ Male ☐ Female
Height	☐ Unknown I__I__I__I cm
Weight	☐ Unknown I__I__I__I kg
BMI	Calculated automatically
Occupation / Job	☐ Metal worker ☐ Roof worker ☐ Other; …… ☐ Unknown
Smoking Status	☐ Active Smoker ☐ Never Smoked ☐ Quit Smoking ☐ Unknown If active smoker or quit smoking: …… packs / year
Co-morbidites	☐ Yes ☐ No, If yes; ☐ Arterial Hypertension ☐ Pulmonary Hypertension; if yes; group; (Can be marked multiple) ☐ Goup I ☐ Group II ☐ Group III ☐ Group IV ☐ Group V ☐ Diabetes mellitus ☐ Congestive Heart Failure ☐ Ischemic heart disease ☐ Neurologic Disease ☐ Malignancy; If yes please specify; ……..

	☐ Liver Disease ☐ Gastroesophageal reflux disease ☐ Long Term Proton Pump Inhibitor Usage ☐ Other; ……..

CONCOMITANT MEDICATIONS (Not Related to CTD / ILD, ongoing medications after diagnosis)

Medication 1 Active Ingredient In Medication	
Start Date / End Date or Ongoing	..... ☐ Ongoing
Indication	……
Medication 2 Active Ingredient In Medication	
Start Date / End Date or Ongoing	..... ☐ Ongoing
Indication	……
Medication 3 Active Ingredient In Medication	
Start Date / End Date or Ongoing	..... ☐ Ongoing
Indication	……
Medication 4 Active Ingredient In Medication	
Start Date / End Date /or Ongoing	..... ☐ Ongoing
Indication	……

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DIAGNOSIS INFORMATION

Date of Diagnosis of CTD	I__I__I / I__I__I / I__I__I__I__I
CTD Type	☐ Rheumatoid Arthritis Joint involvement (0-5 points) ☐ None ☐ 1 large joint ☐ 2-10 large joints ☐ 1-3 small joints (large joints not counted) ☐ 4-10 small joints (large joints not counted) ☐ >10 joints including at least one small joint Serology (at least one test needed for classification; 0-3 points) ☐ None ☐ Negative RF and negative ACPA ☐ Low positive RF or low positive ACPA ☐ High positive RF or high positive ACPA Acute-phase reactants (at least one test needed for classification; 0-1 point) ☐ None ☐ Normal CRP and normal ESR ☐ Abnormal CRP or abnormal ESR Duration of symptoms ☐ None ☐ < 6 weeks ☐ >= 6 weeks SCORE:
	☐ Systemic sclerosis (SSc) ☐ Skin thickening of the fingers of both hands extending proximal to the metacarpophalangeal joints (sufficient criterion) Skin thickening of the fingers (only count the highest score) ☐ None ☐ Puffy fingers ☐ Sclerodactyly of the fingers (distal to MCP but proximal to the PIPs) Finger Tip Lesions ☐ None ☐ Digital Tip Ulcers ☐ Finger Tip Pitting Scars ☐ Telangiectasia Pulmonary arterial hypertension and/or Interstitial Lung Disease (Maximum score is 2) ☐ None ☐ PAH

☐ ILD
☐ PAH and ILD

Scleroderma related antibodies

☐ None
☐ Anti-centromere
☐ Anti-topoisomerasel
☐ Anti-RNA antibody

☐ Abnormal nailfold capillaries
☐ Raynaud's Phenomenon

SCORE:

☐ Mixed connective tissue disease

1. **Serologic criterion: Positive antibodies to U1 RNP antibodies at a titer >= 1:1600**

1. Serologic criterion: Positive antibodies to U1 RNP antibodies at a titer >= 1:1600

☐ Yes ☐ No

2. **Clinical criteria**

1) Edema of the hands	☐ Yes	☐ No
2) Synovitis	☐ Yes	☐ No
3) Myositis	☐ Yes	☐ No
4) Raynaud phenomenon	☐ Yes	☐ No
5) Sclerodactyly	☐ Yes	☐ No

☐ Myositis

Age of onset of first related symptoms

☐ 18-40 ☐ => 40

☐ No biopsy ☐ Biopsy

Muscle Weakness

☐ Objective symmetric weakness, usually progressive, of proximal upper extremities
☐ Objective symmetric weakness, usually progressive, of proximal lower extremities
☐ Neck flexors are relatively weaker than neck extensors
☐ In the legs, proximal muscles are relatively weaker than distal muscles

☐ No biopsy ☐ Biopsy

Skin manifestations

☐ Heliotrope rash
☐ Gottron's papules
☐ Gottron's sign

☐ No biopsy ☐ Biopsy

Other clinical manifestations

☐ Dysphagia or esophageal dysmotility

☐ No biopsy ☐ Biopsy

Laboratory measurements

☐ Anti-Jo-1 (anti-histidyl-tRNA synthetase) autoantibody positivity

☐ Elevated serum levels of creatine kinase (CK)* or lactate dehydrogenase (LDH)* or aspartate aminotransferase (ASAT/AST/SGOT)* or alanine aminotransferase (ALAT/ALT/SGPT)

☐ No biopsy ☐ Biopsy

Muscle biopsy features

☐ Endomysial infiltration of mononuclear cells surrounding, but not invading, myofibres

☐ Perimysial and/or perivascular infiltration of mononuclear cells

☐ Perifascicular atrophy

☐ Rimmed vacuoles

☐ No biopsy ☐ Biopsy

SCORE:

☐ Systemic lupus erythematosus (SLE)

Clinical Domains

Constitutional domain

☐ Fever

Cutaneous domain

☐ Non-scarring alopecia

☐ Oral ulcers

☐ Subacute cutaneous or discoid lupus

☐ Acute cutaneous lupus

Arthritis domain

☐ Synovitis or tendernessin at least 2 joints

Neurologic domain

☐ Delirium

☐ Psychosis

☐ Seizure

Serositis domain

☐ Pleural or pericardial effusion

☐ Acute pericarditis

Hematologic domain

☐ Leukopenia

☐ Thrombocytopenia

☐ Autoimmune hemolysis

Renal domain

☐ Proteinuria> 0.5 g/24 hr

☐ Class Il or V lupus nephritis

☐ Class Ill or IV lupus nephritis

Immunologic Domains

Antiphospholipid antibody domain

☐ Anticardiolipin IgG > 40 GPL 2 or anti-B2GP1 IgG > 40 units or lupus anticoagulant

Complement proteins domain

☐ Low C3 or low C4

☐ Low C3 and low C4

Highly specific antibodies domain

☐ Anti-dsDNA antibody
☐ Anti-Sm antibody

SCORE:

☐ Sjogren's Disease.

☐ Labial salivary gland with focal lymphocytic sialadenitis and focus score of >= 1 foci/4 mm2 (3 points)
☐ Anti-SSA/Ro positive (3 Points)
☐ Ocular Staining Score >= 5 (or van Bijsterveld score >=4) in at least 1 eye (1 Point)
☐ Schirmer's test <= 5 mm/5 minutes in at least 1 eye (1 Point)
☐ Unstimulated whole saliva flow rate <= 0.1 ml/minute (1 Point)

SCORE:

☐ ANCA Associated Vasculitis

The patient followed for a minimum of 3 months.
☐ Yes ☐ No

The patient must be aged>16 years at the time of diagnosis.
☐ Yes ☐ No

Histological proof of vasculitis (including necrotising glomerulonephritis) and/or granuloma formation (granuloma are defined according to the American College of Rheumatology (ACR, 1990) criteria for Wegener's granulomatosis—as histological changes showing granulomatous inflammation within the wall of an artery or in the perivascular or extravascular area (artery or arteriole)3)
☐ Yes ☐ No

Positive serology for ANCA (proteinase 3-ANCA or myeloperoxidase-ANCA; indirect immunofluorescence result alone is acceptable only if ELISA is unavailable in a centre or the diagnosis was made before 1995)
☐ Yes ☐ No

Specific investigations strongly suggestive of vasculitis and/or granuloma (neurophysiology must show mononeuritis multiplex) (from angiography; either magnetic resonance angiography or coeliac axis angiography in PAN), thoracic or neck magnetic resonance imaging/ computed tomography imaging (showing retro-orbital or tracheal disease; neurophysiology) (the presence of IgA in a renal or skin biopsy is suggestive of Henoch Schonlein purpura (HSP). Detection of antiglomerular basement membrane (GBM) antibodies is suggestive of Goodpasture's syndrome. However, we recognise that both IgA deposits and anti-GBM antibodies may occur concurrently with ANCA-positive vasculitis. The exclusion of cases of HSP/Goodpasture's is left to the individual clinician)
☐ Yes ☐ No

Eosinophilia (.10% or .1.56109/l)
☐ Yes ☐ No

Malignancy
☐ Yes ☐ No

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	Infection (including hepatitis B and C, HIV, tuberculosis, subacute bacterial endocarditis) ☐ Yes ☐ No Drugs (including hydralazine, propylthiouracil, cocaine and allopurinol) ☐ Yes ☐ No Secondary vasculitis—rheumatoid arthritis, systemic lupus erythematosus, Sjo¨gren's syndrome, connective tissue disease ☐ Yes ☐ No Behcet's disease, Takayasu's arteritis, giant cell arteritis, Kawasaki's disease, essential mixed cryoglobulinaemia, Henoch Scho¨nlein purpura, anti-GBM disease ☐ Yes ☐ No Vasculitis mimics—for example, cholesterol embolism, calciphylaxis, catastrophic antiphospholipid antibody syndrome, atrial myxoma ☐ Yes ☐ No Sarcoidosis and other non-vasculitic granulomatous disease ☐ Yes ☐ No
Start Date of Symptoms of CTD and first symptom	I__I__I / I__I__I / I__I__I__I__I First symptom: .....
Date of Diagnosis of ILD	I__I__I / I__I__I / I__I__I__I__I
Start Date of Symptoms of ILD	I__I__I / I__I__I / I__I__I__I__I
ILD Symptoms at diagnosis	☐ Symptomatic ☐ Aysmptomatic, If symptomatic; ☐ cough ☐ dyspnea ☐ fatigue ☐ weight loss ☐ other;…….
Extent of lung status:	☐ <20 ☐ >=20
HRCT	HRCT ☐ Done, If Done HRCT Pattern (at CTD-ILD Diagnosis) ☐ Usual Interstitial Pneumonia (UIP) ☐ Non-Specific Interstitial Pneumonia ☐ Organizing Pneumonia ☐ LIP ☐ Unclassified ☐ Not Done ☐ Unknown
6 Min Walking Test (At diagnosis)	☐ Not Done ☐ Done ☐ Not completed ☐ Completed

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	…….meter spO2 (Initial): spO2 (End):
FVC (At diagnosis)	…… mL
FVC - % predicted (At diagnosis)	……
DLCO (At diagnosis)	……. mL/mm Hg/min
DLCO - % predicted (At diagnosis)	
DLCO / VA	.....
Is Lung Biopsy Performed?	☐ No ☐ Unknown ☐ Yes, If Yes Date of Biopsy; ……/……/…….
Biopsy Result	……
ECHO	☐ Done ☐ Not Done ☐ Unknown If Done; Pulmonary Artery Pressure (s): ..... mm/Hg TRV: ..... EF: Pericardial Fluid: ☐ Yes ☐ No
Right Heart Catheterization	☐ Done ☐ Not Done ☐ Unknown If Done; mPAB: PCWP: PVR:
BNP	..... pg/ml
Pro BNP	..... pg/ ml

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LABORATORY PARAMETERS AT DIAGNOSIS

Date of Lab Results	I__I__I / I__I__I / I__I__I__I__I
Antinuclear antibody	.....
Rheumatoid factor	☐ Positive ☐ Negative If Positive: .....
Anti CCP	☐ Positive ☐ Negative If Positive: .....
Creatinine kinase	
CRP (mg/dl)	
Erythrocyte sedimentation rate	
Miyositis specific antibody	☐ Jo-1 ☐ PL-7 ☐ PL-12 ☐ EJ ☐ OJ ☐ Mi-2 ☐ SRP ☐ KS ☐ TIF1γ/α ☐ TIF1β ☐ MJ/NXP-2 ☐ MDA5/CADM-140 ☐ SAE
Anti-ro	☐ Positive ☐ Negative ☐ Unknown
Anti-ro 52	☐ Positive ☐ Negative ☐ Unknown
Anti – ds DNA	.....
Anti-la	☐ Positive ☐ Negative ☐ Unknown
Anti-Scl70	☐ Positive ☐ Negative ☐ Unknown

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Anti-Smith antibody	☐ Positive ☐ Negative ☐ Unknown
Anti rnp	☐ Positive ☐ Negative ☐ Unknown
C - ANCA	☐ Positive ☐ Negative ☐ Unknown
P - ANCA	☐ Positive ☐ Negative ☐ Unknown
Anti PR3	☐ Positive ☐ Negative ☐ Unknown
Anti MPO	☐ Positive ☐ Negative ☐ Unknown

TYPE OF CTD TREATMENT

Type of CTD Treatment Initial Treatment	Start Date: I__I__I / I__I__I / I__I__I__I__I Stop Date: I__I__I / I__I__I / I__I__I__I__I ☐ Ongoing ☐ Corticosteroids; ☐ Low Dose ☐ High Dose ☐ Immunosuppressive Therapy; ☐ Biological immunsupressive (Rituximab); ☐ For CTD ☐ For ILD ☐ Anti-cytokine (tocilizumab); ☐ For CTD ☐ For ILD ☐ CS – DMARD; ☐ For CTD ☐ For ILD ☐ Biological DMARD; ☐ For CTD ☐ For ILD ☐ TS – DMARD; ☐ For CTD ☐ For ILD ☐ Other; …….. ☐ For CTD ☐ For ILD Is Medication change needed ? ☐ Yes ☐ No

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Type of CTD Treatment Follow-up	Reason for the change of previous therapy: ☐ Lack of Efficacy ☐ Intolerance or toxicity; ☐ Related to the lungs ☐ Not related to the lungs; Definition: …… ☐ Physicisan discreation Start Date: I__I__I / I__I__I / I__I__I__I__I Stop Date: I__I__I / I__I__I / I__I__I__I__I ☐ Ongoing ☐ Corticosteroids; ☐ Low Dose ☐ High Dose ☐ Immunosuppressive Therapy; ☐ Biological immunsupressive (Rituximab); ☐ For CTD ☐ For ILD ☐ Anti-cytokine (tocilizumab); ☐ For CTD ☐ For ILD ☐ CS – DMARD; ☐ For CTD ☐ For ILD ☐ Biological DMARD; ☐ For CTD ☐ For ILD ☐ TS – DMARD; ☐ For CTD ☐ For ILD ☐ Other; …….. ☐ For CTD ☐ For ILD
Type of CTD Treatment Follow-up	Start Date: I__I__I / I__I__I / I__I__I__I__I Stop Date: I__I__I / I__I__I / I__I__I__I__I ☐ Ongoing ☐ Corticosteroids; ☐ Low Dose ☐ High Dose ☐ Immunosuppressive Therapy ☐ Biological immunsupressive (Rituximab); ☐ For CTD ☐ For ILD ☐ Anti-cytokine (tocilizumab); ☐ For CTD ☐ For ILD ☐ CS – DMARD; ☐ For CTD ☐ For ILD ☐ Biological DMARD; ☐ For CTD ☐ For ILD ☐ TS – DMARD; ☐ For CTD ☐ For ILD ☐ Other; …….. ☐ For CTD ☐ For ILD

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CLINICAL / LABORATORY PARAMETERS
AT FOLLOW UP IF APPLICABLE (Annually; Including 4th year follow-up)

Symptoms	☐ New symptom ☐ Progression of the symptoms ☐ Regression of the symtoms
Diseases since one year	Infection: ☐ Yes. ☐ None, If yes; ☐ COVID19 Cardiovascular Disease ☐ Yes. ☐ None, If yes; ☐ MI. ☐ Stroke. ☐ Other; …… Malignity ☐ Yes. ☐ None Other Diseases; …….
HRCT (High Resolution Computed Tomography)	☐ Done ☐ Not Done ☐ Unknown If Done; HRCT Pattern (at CTD-ILD Diagnosis) ☐ Usual Interstitial Pneumonia (UIP) ☐ Non-Specific Interstitial Pneumonia ☐ Organizing Pneumonia ☐ LIP ☐ Unclassified
HRCT assesment	☐ Progression ☐ Regression ☐ Stable
HRCT Result	☐ Normal ☐ Abnormal If abnormal;

	☐ Anterior upper lobe ☐ Exuberant honeycombing ☐ Straight edge ☐ Other; …….
6 Min Walking Test (At diagnosis)	☐ Not Done ☐ Done ☐ Not completed ☐ Completed …….meter spO2 (Initial): spO2 (End):
FVC (At follow-up)	…… mL
FVC - % predicted (At follow-up)	……
DLCO (At follow-up)	……. mL/mm Hg/min
DLCO - % predicted (At follow-up)	
DLCO / VA	.....
ECHO	☐ Done ☐ Not Done ☐ Unknown If Done; Pulmonary Artery Pressure (s): ..... mm/Hg TRV: ..... EF: Pericardial Fluid: ☐ Yes ☐ No
Date of Lab Results	I__I__I / I__I__I / I__I__I__I__I
Antinuclear antibody	.....

Rheumatoid factor	☐ Positive ☐ Negative If Positive: .....
Anti CCP	☐ Positive ☐ Negative If Positive: .....
Creatinine kinase	.....
CRP (mg/dl)	.....
Erythrocyte sedimentation rate	.....
Miyositis specific antibody	☐ Jo-1 ☐ PL-7 ☐ PL-12 ☐ EJ ☐ OJ ☐ Mi-2 ☐ SRP ☐ KS ☐ TIF1γ/α ☐ TIF1β ☐ MJ/NXP-2 ☐ MDA5/CADM-140 ☐ SAE
Have there been any changes in the laboratory parameters below compared to the previous follow-up?	☐ Yes ☐ No
Anti-ro	☐ Positive ☐ Negative ☐ Unknown
Anti-ro 52	☐ Positive ☐ Negative ☐ Unknown
Anti – ds DNA	.....
Anti-la	☐ Positive ☐ Negative ☐ Unknown
Anti-Scl70	☐ Positive ☐ Negative ☐ Unknown
Anti-Smith antibody	☐ Positive ☐ Negative ☐ Unknown
Anti rnp	☐ Positive ☐ Negative ☐ Unknown
C - ANCA	☐ Positive ☐ Negative ☐ Unknown

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P - ANCA	☐ Positive ☐ Negative ☐ Unknown
Anti PR3	☐ Positive ☐ Negative ☐ Unknown
Anti MPO	☐ Positive ☐ Negative ☐ Unknown

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END OF STUDY PAGE

Is patient still followed up at the site?

☐ Yes ☐ No

If Yes;

Last Contact Date I__I__I / I__I__I / I__I__I__I__I

If No;

Last Contact Date I__I__I / I__I__I / I__I__I__I__I

Reason:

☐ Lost to follow-up
☐ Death;
Date of death: I__I__I / I__I__I / I__I__I__I__I
Reason for death:.....
☐ Other (.....)