**1. Patient data / Patient personal details**

**Insurance number:…………………………..**

**Name:**…………………………………………

**Date of birth:**…………………………………….

**Contact number:………………………………..**

**Gender:** Female / Male

**Ethnicity/Race:** White / Black / Indian / Asian / Other: …….

**Date of questioning**: ………………………………(year/month/day)

**2. Anamnesis / Details from the medical history**

# 2.1 Lifestyle

**Smoking**: yes / no

if yes: amount (cigarettes/day):…………………

For how many years? …………………………

if not:

Did you smoke earlier? yes / no

if yes: amount (pcs/occasion):…………………………………

For how many years?………………………………………

How long ago did you stop smoking? ……………………………….

**Alcohol consumption**: yes / no

if yes: frequency: occasionally/monthly/weekly/daily

amount (g/day):…………………………………

since when? (years):………………………….

 Alcohol consumption in the last 2 weeks: …………………..

if not:

Did you drink alcohol earlier? yes / no

if yes: frequency: occasionally/monthly/weekly/daily

amount (g/occasion):…………………………………

For how many years?………………………………………

How long ago did you stop drinking alcohol?..........................

 *Guide for estimation of the amount:*

*1 dl beer (4.5 vol. %) = ~3.5 g alcohol*

 *1 dl wine (12.5 vol. %) = ~10 g alcohol*

 *1 dl hard drink (50 vol. %) = ~40 g alcohol*[T1]

 **Diet**: yes / no

 if yes: diabetic / gluten-free / vegetarian / paleolit / lactose-free / other:………………

**Physical activity**: none / occasionally / regularly / intensely

*None: no exercise or exercise for < 3 hours/week for < 2 years*

*Occasionally: < 3 hours/week for >= 2 years*

*Regularly: > 3 hours/week for >= 2 years*

*Intensely: > 7 hours/week for>= 2 years*

**Drug abuse**: yes / no *Prescribed medication should not be included here.*

if yes: type of drug:……………………………………. amount:………………………………………….

for how many years:……………………………

**Job:**  blue collar worker / white collar worker

mining / sewing / other works with long-term vibration exposure

# 2.2 Comorbidities

**Diabetes mellitus**: yes/no

if yes: type I. / type II / type III. / MODY

**Allergies:** yes / no

if yes: specify: inhalative / other nutrition / drug / eczema / other

**Hypertension:** yes / no

 if yes: since (year):

**Malignancy in history:** yes / no

 if yes: date: ……………….(year/month/day)

 please specify: …………………..

**Thromboembolic events in history:** yes / no

 if yes: deep vein thrombosis: yes / no

 if yes: date:…………….(year/month/day)
 (többször hozzáadható)

pulmonary embolism: yes / no

 if yes: date:…………….(year/month/day)
 (többször hozzáadható)

 repeated miscarriages or stillbirths: yes / no

 if yes: date:…………….(year/month/day)
 (többször hozzáadható)

 stroke: yes / no

 if yes: date:…………….(year/month/day)
 (többször hozzáadható)

 myocardial infarction: yes / no

 if yes: date:…………….(year/month/day)
 (többször hozzáadható)

**Autoimmunity in history:** yes /no

if yes: systemic / organ specific

if systemic: Rheumatoid arthritis / Systemic lupus erythematosus / Scleroderma / Sjögren's Syndrome / Antiphospholipid syndrome / Idiopathic inflammatory myopathies / Vasculitis / Other

if Rheumatoid arthritis: date of diagnosis: ……..(year/month/day)

if Systemic lupus erythematosus:date of diagnosis:…..(year/month/day)

if Scleroderma: date of diagnosis: ……..(year/month/day)

if Sjögren's Syndrome: date of diagnosis: ……..(year/month/day)

if Antiphospholipid syndrome: date of diagnosis:....(year/month/day)

if Idiopathic inflammatory myopathies: date of diagnosis:......(year/month/day)

if Vasculitis: date of diagnosis: ……..(year/month/day)

if Other:please specify: …… date of diagnosis: ……..(year/month/day)

if organ specific: Type I diabetes / Hashimoto's thyroiditis / Graves' disease / Addison’s disease / Autoimmune hepatitis / Primary biliary cholangitis / Primary sclerosing cholangitis / Multi-glandular diseases / Autoimmune pancreatitis / Coeliac disease / Autoimmune hemolytic anemia / Autoimmune thrombocytopenic purpura / Myasthenia gravis / Multiple sclerosis / Inflammatory neuropathies / Other

if Type I diabetes: date of diagnosis: ……..(year/month/day)

if Hashimoto's thyroiditis: date of diagnosis: ……..(year/month/day)

if Graves' disease: date of diagnosis: ……..(year/month/day)

if Addison’s disease: date of diagnosis: ……..(year/month/day)

if Autoimmune hepatitis: date of diagnosis: ……..(year/month/day)

if Primary biliary cholangitis: date of diagnosis: ……..(year/month/day)

if Primary sclerosing cholangitis:date of diagnosis: ….(year/month/day)

if Multi-glandular diseases: date of diagnosis: ……..(year/month/day)

if Autoimmune pancreatitis: date of diagnosis: ……..(year/month/day)

if Coeliac disease: date of diagnosis: ……..(year/month/day)

if Autoimmune hemolytic anemia: date of diagnosis:...(year/month/day)

if Autoimmune thrombocytopenic purpura:date of diagnosis:....(year/month/day)

if Myasthenia gravis: date of diagnosis: ……..(year/month/day)

if Multiple sclerosis: date of diagnosis: ……..(year/month/day)

if Inflammatory neuropathies: date of diagnosis: ….(year/month/day)

if Other: please specify: ……………

 date of diagnosis:.......(year/month/day)

**Raynaud phenomenon:** yes / no

if yes: start date: …….. (year)

**Date of first non-Raynaud symptom:** ……. (year)

**Disease duration** **(year)**: ………… \*

*\* Calculated from the first non-Raynaud symptom.*

**Scleroderma subgroup:** limited cutaneous SSc / diffuse cutaneous SSc/ SSc sine scleroderma (based on medical documentations)

**Intestinal lung disease:** yes / no \*

 if yes: date of diagnosis: ……………….(year/month/day)

*\* Definition: Fibrosis proven by HRCT and ( <80% FVC or <80 DLCO) on pulmonary function tests.*

**Ischemic heart disease\*:** yes / no

 if yes: date of diagnosis: ……………….(year/month/day)

*\* Definition: previews myocardial infarct or PTCA or CABG or pozitiv CT-angiography or pozitiv coronary angiography*

**Congestive heart disease\*:** yes / no

*\* Definition: using of loop diuretics*

**Scleroderma renal crisis:** yes / no

Renal replacement therapy (dialysis) due to SRC: yes / no

**Malabsorption**: yes / no

*Malabsorption definition: decreased serum protein / nutritional supplement / long-term weight loss.*

**Bacterial overgrowth:** yes / no

*Bacterial overgrowth definition: proven by H2 exhalation / cyclic antibiotic treatment.*

**Hiperalimentation required:** yes / no

**Asthma:**  yes / no

 if yes: date of start of treatment: …………….(year/month/day)

**COPD:** yes / no

 if yes: date of start of treatment: …………….(year/month/day)

**Gout in history:** yes / no

 if yes: date of first attack: ………….(year/month/day)

**Spondyloarthritis in history:** yes / no

 if yes: date of diagnosis: ………….(year/month/day)

axial spondyloarthritis / peripheral spondyloarthritis / reactive arthritis / psoriatic arthritis / enteropathic arthritis / endogenous uveitis

**Other internal organ disease**: yes / no

 if yes: heart / lung / gastrointestinal / liver / renal / hematologic / thyroid

 please specify: …………….

**Long lasting / recurrent fever of unknown origin:** yes / no

 if yes: date of start of the first episode: ………….(year/month/day)

 date of last episode: ………….(year/month/day)

**Unexplained weight loss in history:** yes / no

 if yes: date of start of the weight loss: ………….(year/month/day)

 body weight at this time (kg):……………

 date of end of the weight loss: ………….(year/month/day)

 body weight at this time (kg):…………….

 weight loss of >10% in the last 12 months: yes / no

**Osteoporosis (DEXA):** yes / no

 if yes: date of diagnosis: ………….(year/month/day)

 any osteoporotic fracture: yes / no

 if yes date of the first fracture:………….(year/month/day)

 Number of the osteoporotic fractures:…………..

**Previous digital painful ischemia:** yes / no

**Previous digital ulcer:** yes / no

**Previous digital gangrene:** yes / no

**Lung biopsy:**  yes / no

 if yes: please copy the histology findings: ………………

**Renal biopsy:** yes / no

 if yes: date: ………….(year/month/day)

please specify: ……………….

# 2.3 Family history

**Thromboembolic events in family history**

if yes: relationship to patient:\*...........................

deep vein thrombosis / pulmonary embolism / stroke / myocardial infarction

**Autoimmunity in family:** yes /no

 if yes: relationship to patient\*:…………………………………….

systemic / organ specific

if systemic: Rheumatoid arthritis / Systemic lupus erythematosus / Scleroderma / Sjögren's Syndrome / The antiphospholipid syndrome / Idiopathic inflammatory myopathies / Vasculitis / Other

if Other: please specify: ……………

if organ specific: Type I diabetes / Hashimoto's thyroiditis / Graves' disease / Addison’s disease / Autoimmune hepatitis / Primary biliary cholangitis / Primary sclerosing cholangitis / Multi-glandular diseases / Autoimmune pancreatitis / Coeliac disease / Autoimmune hemolytic anemia / Autoimmune thrombocytopenic purpura / Myasthenia gravis / Multiple sclerosis / inflammatory neuropathies / Other

if Other: please specify: ……………

**Spondyloarthritis in family history**: yes / no

 if yes: relationship to patient\*: …………………………………….

**Asthma in family history:** yes / no

 if yes: relationship to patient\*: …………………………………….

**COPD in family history:** yes / no

 if yes: relationship to patient\*:…………………………………….

\* **relationship:** father / mother / sibling / child / paternal grandfather / paternal grandmother / maternal grandmother / maternal grandfather / paternal cousin / maternal cousin / father sibling (uncle, aunt) / mother sibling (uncle, aunt) / siblings child (nephew, niece) / grandchild / paternal grandfathers sibling / paternal grandmothers sibling / maternal grandfathers sibling / maternal grandmothers sibling / other blood relation / spouse (husband, wife, other not blood relation)

3. Complaints / Risk factors

**Current painful digital ischemia:** yes / no

**Morning stiffness:** yes / no

 if yes: Morning stiffness duration (min): …….

**Sicca symptoms (dry eyes and/or dry mouth): yes / no**

**Esophageal symptoms (dysphagia or reflux):** yes / no

 if yes: dysphagia: yes/ no

 reflux: yes / no

**Stomach symptoms (early satiety, heartburn, vomiting):** yes / no

 if yes: early satiety: yes / no

 heartburn: yes / no

 vomiting: yes / no

**Intestinal symptoms (diarrhea, bloating, constipation):** yes /no

 if yes: diarrhea: yes / no

 bloating: yes / no

 constipation: yes / no

**(Pseudo-)obstruction (vomiting or constipation, with dilatation of bowels on imaging):** yes/no

**Worsening of cardiopulmonary symptoms within last month:** yes / no

**Worsening of Raynaud symptoms within last month: yes / no**

**Worsening of skin thickness within last month: yes / no**

**Palpitations:** yes / no

**Dyspnoe:** yes / no

**Exertional dyspnoe:** yes / no

**Exposure to solvents:**  yes / no

4. Questionnaires

**HAQ score:** ………

**SSc-HAQ - Raynaud complaint (0-100):** ……….

**SSc-HAQ - Digital ulcer complaints (0-100):** ……….

**SSc-HAQ - Gastrointestinal complaints (0-100):** ………

**SSc-HAQ - Lung complaints (0-100):** ……..

**SSc-HAQ – Overall (0-100):** …..

**Pain in the last week (0-100):** …….

**Joint pain in the last week (0-100):** ………..

**Fatigue in the last week (0-100):** ………

**Arthritis activity in the last week (0-100):** ………

Skin thickness questionnaire

**SF-36 PCS (%):** ………

**SF-36 MCS (%):** ……….

**UCLA:** ………..

5. Status

**Body weight (kg):** …..………………………

**Body height (cm):** ………………

**BMI** (automatikusan számolt)

**Blood pressure (Hgmm):** ……..… / ………

**Heart rate:** ……………………… /minute

**6MWT:** yes / no

if yes: O2 saturation at rest: ………%

Blood pressure at rest ……………Hgmm

Heart rate at rest ………………../min

O2 saturation at exercise: ……….%

Blood pressure at exercise……………Hgmm

Heart rate at exercise………………../min

Distance on 6MWT (m): …………

Interruption 6MWT: yes / no

if yes: Reason for interruption: cardiological / musculoskeletal / other

**Borg scale (0-10):** …… \*

**NYHA:** 1 / 2 / 3 / 4 \*\*

*\*Instruction for Borg Dyspnea Scale*

*Use this scale to rate the difficulty of breathing. It starts at number 0 where breathing is causing no difficulty at all and progresses through to number 10 where breathing difficulty is maximal.*

*0 – Nothing at all*

*1 – Very slight*

*2 – Slight*

*3 – Moderate*

*4 – Somewhat severe*

*5 – Severe*

*6*

*7 – Very severe*

*8*

*9 – Very, very severe (almost maximal)*

*10 – Maximal*

*\*\*NYHA Classification*

*1 – No symptoms and no limitation in ordinary physical activity.*

*2 – Mild symptoms (mild shortness of breath and/or angina) and slight limitation during ordinary activity.*

*3 – Marked limitation in activity due to symptoms, even during less-than-ordinary activity (walking short distances 20-100 m). Comfortable only at rest.*

*4 – Severe limitation. Experiences symptoms even while at rest. Mostly bedbound patients.*

6. Physical examination

**Scleroedema:**  yes/no

**Sclerodactyly:** yes / no

**Puffy fingers (current or previous):** yes / no

**Proximal scleroderma:** yes / no

**Scleroderma subgroup (based on current physical examination):** limited cutaneous SSc / diffuse cutaneous SSc / SSc sine scleroderma

**Pitting scars on fingertips:** previously / current / never

**Gangrene:** previously / current / never

**Digital ulcer:** previously / current / never

**Telangiectasia:** yes / no

**Pigmentation disturbances:** yes / no

**Muscle weakness\*:** yes / no

**Muscle atrophy:** yes / no

**Subcutaneous calcinosis:**  previously / current / never

**Calcinosis complicated by infection or requiring surgery**: yes / no

**Disease activity by physician (0-100):** ………….

**Joint activity by physician (0-100):** ………….

**Musculoskeletal activity by physician (0-100):** ………….

**Overall disease severity by physician (0-100):…………**

**Presence of anasarca: yes / no**

\*Proximal muscle weakness on clinical examination defined as shoulder abduction and/or hip or knee flexion less than 5/5 power (not due to contracture or pain)

**Tenderness joints:** yes / no

 if yes:

 right IP: yes / no

 right PIP II: yes / no

 right PIP III: yes / no

 right PIP IV: yes / no

 right PIP V: yes / no

 right MCP I: yes / no

 right MCP II: yes / no

 right MCP III: yes / no

 right MCP IV: yes / no

 right MCP V: yes / no

 right Wrist: yes / no

 right Elbow: yes / no

 right Shoulder: yes / no

 right Knee: yes / no

 left IP: yes / no

 left PIP II: yes / no

 left PIP III: yes / no

 left PIP IV: yes / no

 left PIP V: yes / no

 left MCP I: yes / no

 left MCP II: yes / no

 left MCP III: yes / no

 left MCP IV: yes / no

 left MCP V: yes / no

 left Wrist: yes / no

 left Elbow: yes / no

 left Shoulder: yes / no

 left Knee: yes / no

Number of tenderness joints: ……

**Swollen joints:** yes / no

 if yes:

 right IP: yes / no

 right PIP II: yes / no

 right PIP III: yes / no

 right PIP IV: yes / no

 right PIP V: yes / no

 right MCP I: yes / no

 right MCP II: yes / no

 right MCP III: yes / no

 right MCP IV: yes / no

 right MCP V: yes / no

 right Wrist: yes / no

 right Elbow: yes / no

 right Shoulder: yes / no

 right Knee: yes / no

 left IP: yes / no

 left PIP II: yes / no

 left PIP III: yes / no

 left PIP IV: yes / no

 left PIP V: yes / no

 left MCP I: yes / no

 left MCP II: yes / no

 left MCP III: yes / no

 left MCP IV: yes / no

 left MCP V: yes / no

 left Wrist: yes / no

 left Elbow: yes / no

 left Shoulder: yes / no

 left Knee: yes / no

Number of swollen joints: ……

**DAS28 (ESR):** ………….

**DAS28 (CRP):** ………….

**SDAI:** …………

**CDAI:** ………….

**Contracture:** yes / no

 if yes, small joint contracture: yes / no

 if yes: number of small joints with contracture: ……

 large joint contracture\*: yes / no

 if yes: number of large joints with contracture: ……

**Tendon friction rubs:** yes / no

\*Joint contracture defined as any degree of contracture with the inability to reduce the joint to the anatomically neutral position in the large joints, specifically elbows and knees.

**Modified Rodnan skin score:** yes / no

 if patient has MRSS: face: 0 / 1 / 2 / 3

 chest: 0 / 1 / 2 / 3

 stomach: 0 / 1 / 2 / 3

 right digits: 0 / 1 / 2 / 3

 right hand: 0 / 1 / 2 / 3

 right lower arm: 0 / 1 / 2 / 3

 right upper arm: 0 / 1 / 2 / 3

 right thigh: 0 / 1 / 2 / 3

 right foot: 0 / 1 / 2 / 3

left digits: 0 / 1 / 2 / 3

 left hand: 0 / 1 / 2 / 3

 left lower arm: 0 / 1 / 2 / 3

 left upper arm: 0 / 1 / 2 / 3

 left thigh: 0 / 1 / 2 / 3

 left foot: 0 / 1 / 2 / 3

 Modified Rodnan skin score: ………..

7. Examinations

# 7.1 Laboratory / immunserology\*

**Date of laboratory examination:** ………….. (year/month/day)

|  |  |
| --- | --- |
| **Erythrocyte sedimentation rate (mm/h)** |  |
| **CRP (mg/l)** |  |
| **Hemoglobin (g/l)** |  |
| **Hematokrit (%)** |  |
| **RDW (%CV)** |  |
| **MCV (fl)** |  |
| **Iron (g/dl)** |  |
| **Leukocyte /ul** |  |
| **Lymphocyte /ul** |  |
| **Platelets /ul** |  |
| **MPV (fl)** |  |
| **Urea (mmol/l)** |  |
| **Creatinine (umol/l)** |  |
| **GFR (ml/min/1.73m2)** |  |
| **Uric acid (umol/l)** |  |
| **LDH (U/l)** |  |
| **Total protein (g/l)** |  |
| **Albumin (g/l)** |  |
| **C3 (g/l)** |  |
| **C4 (g/l)** |  |
| **Creatine-kinase (U/l)** |  |
| **Amilase U/ml** |  |
| **Lipase U/ml** |  |
| **TSH mU/l** |  |
| **HgbA1c (%)** |  |
| **Total cholesterol mmol/l** |  |
| **HDL cholesterol mmol/l** |  |
| **Serum triglycerides mmol/l** |  |

**Hypocomplementaemia:** yes / no

**Proteinuria (>300mg/dl):** yes / no

**Haematuria:** yes / no

if yes: renal hematuria / extarenal hematuria

**Blood type:** A / B / AB / 0

Rh positive / Rh negative

**Direct Coombs test:**  yes / no *\**

 if yes: Date: ………….. (year/month/day)

positive / negative

**Cryoglobulinemia**: yes / no *\**

 if yes: Date: ………….. (year/month/day)

positive / negative

**Serum electrophoresis:** yes / no *\**

 if yes: Date: ………….. (year/month/day)

Percent of gamma zone (%): …………

Paraproteins: yes / no

 if yes: Percent of paratproteins (%): ………..

 Please specify the type of paraproteins: ……………….

**Immunoglobulins:** yes /no *\**

 if yes: Date: ………….. (year/month/day)

 IgG (g/l):………….

IgM (g/l): ………..

IgA (g/l):……….

Total IgE (U/ml):………….

**Immunserology:**  yes / no

if yes: ANA: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

ACA: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Scl-70: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

RNS pol III: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

anti-Pm-Scl: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

anti-Ku: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

anti-Th/To: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Fibrillarin: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Ro-52: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

NOR90: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

 Jo-1: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

PL-7: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

PL-12: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Mi-2: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

 SRP: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

 RIB: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

 AMA-M2 (PDH): positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

TIF1y: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

MDA5: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

NXP2: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

SAE1: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

EJ: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

OJ: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Anti-SSA: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Anti-SSB: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Anti-CCP: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

RF: positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

TPO positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

TG positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

ANCA positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

dsDNS positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

M2 positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

LKM1 positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

SLA positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

SMA positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Sp100 positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Transglutaminase positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Endomysium positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Gliadin positive / negative / unknown

 if positive or negative:Date: ………….. (year/month/day)

Any other positive autoantibody: yes / no

if yes: Specify: ………..

 Date: ………………(year/month/day)

(többször hozzáadható)

*\*Results within 3 months of the time of examination may be accepted.*

# 7.2. Imaging techniques

**Esophagus X-ray**\***:** yes / no

 if yes, Date: ………….. (year/month/day)

 Esophageal dysmotility: yes / no

 Esophageal stricture: yes / no

 Esophageal dilatation: yes / no

\**Results within 3 months of the time of examination may be accepted.*

**Lung X-ray**\***:** yes / no\*

 if yes, Date: ………….. (year/month/day)

 Any fibrosis: yes / no

 if yes: Basal fibrosis: yes / no

 Diffuse fibrosis: yes / no

\**Results within 3 months of the time of examination may be accepted.*

**Lung HRCT**\***:**  yes / no \*

 if yes, Date: ………….. (year/month/day)

 Basal fibrosis: yes / no

 Diffuse fibrosis: yes / no

 Fibrosis % involvement: <20% / intermediate / >20% / unknown

 Any ground glass opacity: yes / no

 Any hone combing: yes / no

 Any reticular changes: yes / no

 Any tractions: yes / no

 Any bullae: yes / no

 Any lymp node enlargement: yes / no

 Any granuloma yes / no

 Any air trapping yes / no

 Any consolidation yes / no

 Any emphysema yes / no

 Any pleural effusion yes / no

Specify the pattern of fibrosis: usual interstitial pneumonia (UIP) / non-specific interstitial pneumonia (NSIP) / organising pneumonia (OP) / diffuse alveolar damage (DAD) / respiratory bronchiolitis / desquamative interstitial pneumonia / lymphoid interstitial pneumonia / unknown

Any other suspected symptom on HRCT then fibrosis: yes / no

if yes: infection: yes / no

 if yes: bronchoscopy performed: yes / no

 if yes: please insert finding: ……

 microbiology performed on ……date…

 if yes: please insert finding: ……

if yes: malignancy: yes / no

 if yes: bronchoscopy performed: yes / no

 if yes: please insert finding: ……

 microbiology performed: yes / no

 if yes: please insert finding: ……

 biopsy / histology performed: yes / no

 if yes: please insert finding: ……

Insert the complete HRCT finding, please: ………………………………..

\**Results within 6 months of the time of examination may be accepted.*

**Esophagogastroscopy**\***:** yes / no \*

 if yes, Date: ………….. (year/month/day)

 Gastroesophageal reflux: yes / no

 Barrett’s oesophagus: yes / no

 Oesophagus stricture: yes / no

 Other oesophagus abnormality: yes /no

 if yes: please define: ………………

 Gastric ulcer: yes /no

Vascular ectasia: yes /no

 Bleeding / haemorrhage: yes /no

 Watermelon stomach: yes / no

 Other gastric abnormality: yes / no

 if yes: please define: ………………

 Duodenal ulcer: yes /no

 Celiac disease (scalloped duodenal folds, grooves and fissurations): yes /no

 if yes: confirmed by histology: yes / no

\**Results within 3 months of the time of examination may be accepted.*

**Colonoscopy**\***:**  yes /no \*

 if yes, Date: ………….. (year/month/day)

 Polyp: yes / no

 if yes: Tubular /Villous6 Sessile /Serrated

 Diverticulosis: yes /no

Vascular ectasia: yes /no

Hemorrhoides: yes / no

 if yes: external /internal

 Bleeding / haemorrhage: yes /no

Other colon abnormality: yes /no

 if yes: please define: ………….

\**Results within 3 months of the time of examination may be accepted.*

# 7.3. Electrocardiography / echocardiography / right heart catheterization / spiroergometry

**Electrocardiography**\***:**  yes / no \*

 if yes: Date: ………….. (year/month/day)

Rhythm: Sinus rhythm/ Atrial fibrillation/ Atrial flutter/ Atrial paced/ AV paced / Other

 Atrioventricular block: 1st degree/ 2nd degree/ 3rd degree/ no

Bundle branch block: No/ Incomplete RBBB/ RBBB/ Incomplete LBBB/ LBBB/

other

Syncope secondary to conduction blocks: yes / no

 Arrythmias requiring therapy: yes / no

 Atrial arrhythmias requiring electric cardioversion: yes / no

 Syncope secondary to conduction blocks: yes / no

 Right axis deviation: yes / no

 Ventricular arrhythmias: yes / no

 Atrial arrhythmias: yes / no

 *\* Current hospitalization or outpatient visit*

**Holter-ECG**\***:** yes / no \*

 if yes, Date: ………….. (year/month/day)

maximum / minimum / average heart rate

Asystolia longer than 2 sec: yes / no

Ischemic period: yes / no

Arrhythmias: yes / no

 if yes: ventricular / supraventricular

Total number of premature atrial complexes: ………..

Total premature ventricular complexes: ………..

*\*Results within 3 months of the time of examination may be accepted.*

**Pacemaker:** yes /no

 Pacemaker implanted: yes / no

If Yes: Type: Temporary / permanent

Date:.................................

Reason for pacemaker: Bradyarrhythmia / Treatment of left ventricular outflow tract obstruction / Cardiac resynchronisation therapy / Sudden cardiac death / AV block / Other

Type of pacemaker: AAI / VVI / DDD / ICD / CRT-P / CRT-D / CCM

**Echocardiography\*:**  yes / no

 if yes: Date: ………….. (year/month/day)

 Pericardial effusion: yes / no

 if yes, small / moderate / large *\*\**

 Calculated PAPsys (mmHg): …………

 Pulmonary hypertension\*\*\*\*: yes / no

 Left ventricular ejection fraction (%): …………

 Tricuspid Annular Plane Systolic Excursion (TAPSE) (mm): ……

 Severe right ventricular dysfunction (TAPSE≤14): yes / no

 Diastolic disorder\*\*\*: yes / no

 E (cm/sec): ………….

A (cm/sec): ………….

 e’ (average of septal and lateral) (cm/sec):

 E/e’<8 / 8<E/e’<14 / E/e’>14

 Prosthetic valve: yes / no

 if yes: mitral / aortic / tricuspid / pulmonary

 Left ventricular hypertrophy\*\*\*\*\*: yes / no

Septal thickness (cm):…………..

Posterior wall thickness (cm):……………

Left ventricular enddiastolic diameter (cm):……………..

 Calculated heart weight (g):………………………

 Heart weight/body surface area:…………… (g/m2)

 Any enlarged chamber: yes / no

 if yes:

 left ventricle: yes / no

 left atrium: yes / no

 right ventricle: yes / no

 left atrium: yes / no

 Significant heart valve disease: yes / no

 Any sign of left heart dysfunction/failure: ………..yes / no

 Insert the complete echocardiography finding, please:…………..

*\*Results within 3 months of the time of examination may be accepted.*

*\*\*small: localized or < 10 mm*

*moderate: 10-20 mm*

*large: > 20 mm*

*\*\*\*: impaired relaxation or pseudonormal mitral inflow or restrictive mitral inflow*

*\*\*\*\*: Calculated PAPsys (mmHhmm) ≥ 40*

**Right heart catheterization:** yes / no *\**

 if yes:

date of catheterization:………………… (year/month/day)

Pulmonary hypertension: yes / no

 Pulmonary arterial systolic pressure (mmHg): …………

 PAP distolic (mmHg): …………

 PAP mean (mmHg): …………

 Pulmonary capillary wedge pressure (PCWP) (mmHg): …………

 Pulmonary arterial hypertension if PAP mean ≥ 25 and PCWP ≤ 15 Hgmm

 Pulmonary resistance (dyn/s/cm-5)\*: ………..

 Cardiac index (l/min/m2): ………….

 Cardiac output (l/min): ……….

 Vasodilatator responder: yes / no

**Coronarography:** yes / no *\**

 if yes:

Date: ………….. (year/month/day)

 Significant stenosis: yes / no

 if yes: LM / LAD / LCX / RCA

 Stent implantation: yes / no

 if yes: number of stents: 1 / 2 / 3

 Indication for surgery intervention: yes / no

**Exercise electrocardiogram:** yes / no *\**

 if yes: Date: ………….. (year/month/day)

 Ischemic event: yes / no

**Carotid ultrasonography:** yes / no *\**

 if yes: Date: ………….. (year/month/day)

 Atherosclerotic plaque: yes / no

Significant stenosis: yes /no

# 7.4 Pulmonary function test

**Pulmonary function test\*:** yes / no

 if yes, Date: ………….. (year/month/day)

 FVC (%): …………

 FEV1 (%): …………

 VC (%): ………..

 TLC (%): ……….

 FRC (%): ………..

 TLCO (%): …………

 KCO (%): ………..

 RV (%):………….

 R (kPa/L/s):……………

 FEV1/FVC: …………

*\*Results within 3 months of the time of examination may be accepted.*

# 7.5 Capillary microscopy

**Capillary microscopy***\****:** yes / no

 if yes: Date: ………….. (year/month/day)

 SSc pattern: yes / no

 if yes: Pattern: early / active / late

 Decreased capillary density*\*\**: yes / no

*\*Results within 3 months of the time of examination may be accepted.*

*\*\*Decreased capillary density: less than 7 loops/mm.*

# 7.6 Bronchoalveolar lavage\*: yes / no *\**

 if yes: Date: ………….. (year/month/day)

Total volume (ml): …………

Cell count /ml: …………….

Macrophages (%): ………..

Lymphocytes (%): …………

Neutrophils (%): ………

Eosinophils (%): ………..

Basophils (%): ………..

Plasma cells (%): ……….

Immunohistochemistry or flow cytometry: yes / no

 if yes: Add the complete findings:……………………

Microbiology: yes / no

 if yes: Add the complete findings:……………………

*\*Results within 3 months of the time of examination may be accepted.*

8. Scores

**Damage score**: (calculated from previously given data):……………………..





Modified activity Score (calculated from previous data)

EScSG activity score (calculated from previous data)

New Activity score (calculated from previous data)

9. Therapy

**Prednisolone /methylprednisolone:** yes / no

if yes: dose (mg/day): ………….

 start date: ………….. (year/month/day)

 stop date: ………….. (year/month/day)

**NSAID:** yes / no

 if yes: occasionally / regularly

 daily: yes / no

 weekly : yes / no

**Cyclophosphamide:**  yes / no

if yes: dose (mg/administration): ………….

 start date: ………….. (year/month/day)

 stop date: ………….. (year/month/day)

route of administration: oral / intravenous

**Methotrexate:**  yes / no

if yes: dose (mg/week): ………….

 start date: ………….. (year/month/day)

 stop date: ………….. (year/month/day)

route of administration: oral / subcutaneous

**Azathioprine:**  yes / no

if yes: dose (mg/day): ………….

 start date: ………….. (year/month/day)

 stop date: ………….. (year/month/day)

**Mycophenolate mofetil:** yes / no

if yes: dose (g/day): ………….

 start date: ………….. (year/month/day)

 stop date: ………….. (year/month/day)

**Rituximab:** yes / no

**Tnf alpha antagonist:** yes / no

**Autologus stem cell transplantation:** yes / no

**Immunglobulins:** yes / no

**Tocilizumab:** yes / no

**Abatacept:** yes / no

**JAK inhibitors:** yes / no

**Bosentan:** yes / no

**Ambrisentan:** yes / no

**Macitentan:**  yes / no

**Sildenafil:** yes / no

**Vardenafil:** yes / no

**Tadalafil:** yes / no

**Riociguat:** yes / no

**Prostanoids:**  yes / no

**Ca channel blockers:** yes / no

**Pentoxyphyllin:** yes / no

**ACE inhibitors:** yes / no

**Angiotensin II receptor blockers (ARBs):** yes / no

**Proton pump inhibitors:** yes / no

**H2 receptor blockers:** yes / no

**Prokinetics:**  yes / no

**Diuretics:** yes / no

**Anti-platelet agent:**  yes / no

**Pirfenidon:** yes / no

**Nintedanib:**  yes / no

**Oxygen supply:** yes / no

**Vitamin-D:** yes / no

**Calcium:** yes / no

**Thyroid hormone substitution:**  yes /no

**Bronchodilator treatment:** yes / no

if yes: Adrenergic inhalative agents: yes / no

if yes: alpha and beta adrenergic receptor agonists / non selective beta adrenergic receptor agonists / selective beta-2 adrenergic receptor agonists / combination of adrenergic agents / combination of adrenergic with corticosteroids or others except anticholinergic agents / combination of adrenergic anticholinergic agents

Other inhalative agents: yes / no

if yes: glucocorticoids / anticholinergic agents / anti-allergic agents other than corticosteroids (chromoglic acid or nedocromil)

Systemic adrenergic agents: yes / no

if yes: alpha and beta adrenergic receptor agonists / non selective beta adrenergic receptor agonists / selective beta-2 adrenergic receptor agonists

Other systemic agents: yes / no

if yes: xantins / xantins with adrenergic agents / leucotriene-receptor antagonists / other agents

**Other, specify please**:…………………………..

9. Epicrisis

**Multidisciplinary (ILD) team discussion**: yes / no
Who was present: Surgeon / Internist / Oncologist /Pathologist / Radiologist/Cardiologist/Pulmonologist

**Type of discharge**

To home/To another inpatient ward/ The patient died/ other

**Planned next intervention:** date (dd/mm/yyyy): ………./………./……….

**Comments**:.................

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