

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

Dear colleagues,

Thank you very much for accepting our invitation for this international survey on the diagnosis and treatment of acute exacerbations of IPF - a constant challenge in the treatment of patients with IPF.

We believe that your feedback will help all of us to establish future, international projects in order to improve the diagnosis and treatment of this detrimental complication.

On behalf of the AE-IPF team within “The IPFProject”

Kind regards,

Michael Kreuter

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

Introductory questions to the participants:

Where are you from?

If you want to remain anonymous proceed to the next question.

Where is your working place?

- ☐ Specialised ILD center / university center
- ☐ Pneumology department (non-university center)
- ☐ Intensive care unit
- ☐ Other

How many IPF patients do you have in your practice or center?

0 400 800

How many AE-IPFs do you see per year?

0 200 400

What is your name?

(and E-mail: voluntary, in case you want to be contacted for further projects of the "IPFproject")

If you want to remain anonymous proceed to the next question.

Name:

E-Mail:

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

What is in your experience the average time of symptoms of AE-IPF before the patient initially contacts a physician?

☐ 24 hours

☐ 3 days

☐ 1 week

☐ 2 weeks

☐ > 2-4 weeks

☐ > 4 weeks

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Who do most patients with AE-IPF initially contact?

- ☐ General practitioner (GP)
- ☐ Another specialist (e.g. internal medicine, rheumatology)
- ☐ A non-ILD specialized pulmonologist
- ☐ Other options
- ☐ A community clinic without specialized ILD center (including emergency department)
- ☐ A specialized ILD center

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Where should patients with an AE-IPF be hospitalized ideally if hospitalization is considered necessary?

- ☐ A unit of interstitial lung diseases **with** possibility to be transferred to an intensive care unit (ICU)
- ☐ A unit of interstitial lung diseases **without** access to an ICU
- ☐ A general pulmonology department / section **with** possibility to be transferred to ICU
- ☐ A general pulmonology department / section **without** an ICU
- ☐ A department of internal medicine **with** possibility to be transferred to ICU
- ☐ A department of internal medicine **without** an ICU
- ☐ Other options

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Which laboratory tests are in your opinion / experience mandatory to be performed during diagnostic workup of an AE-IPF? **Multiple answers possible**

- ☐ Blood gas analysis
- ☐ Standard laboratory values including CRP
- ☐ In case of elevated CRP always Procalcitonin
- ☐ D-Dimer
- ☐ Troponins
- ☐ NT-proBNP/BNP
- ☐ KL-6
- ☐ Others blood tests:

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Which further diagnostic procedures are in your opinion / experience mandatory to be performed during diagnostic workup of an AE-IPF? **Multiple answers possible**

- | | |
|---|---|
| <input type="checkbox"/> Chest x-ray | <input type="checkbox"/> Bronchoalveolar lavage always |
| <input type="checkbox"/> HRCT / multislice thin-section CT (without contrast media) | <input type="checkbox"/> Bronchoalveolar lavage only if infection is suspected and the patient is in an appropriate condition to undergo bronchoscopy |
| <input type="checkbox"/> CT with contrast media (even in the absence of clinical suspicion of pulmonary embolism) | <input type="checkbox"/> Echocardiography |
| <input type="checkbox"/> Others: | |

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What specimens do you collect regularly for microbiology assessments in suspected AE-IPF? **Multiple answers possible**

☐

Blood

☐

BAL

☐

Sputum

☐

Urine

☐

Induced sputum

☐

None of the above

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Which pathogens do you regularly screen for in AE-IPF? **Multiple answers possible**

- ☐ CMV-PCR/CMV pp65Ag
- ☐ Pneumocystis jiroveci
- ☐ Aspergillus Antigen
- ☐ Candida Antigen
- ☐ Screening for atypical pathogens (Serum and or urine)
- ☐ Interferon gamma release assay (IGRA) for latent tuberculosis
- ☐ Influenza
- ☐ RSV
- ☐ None of the above
- ☐ Others (e.g. other viruses):

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Do you treat AE-IPF with immunomodulation / corticosteroids? **Multiple answers possible**

- ☐ Prednisolone 1 mg / kg / day, followed by slow tapering (over weeks)
- ☐ Methylprednisolone or equivalent 500 mg-1000 mg / day for 3 days, followed by slow tapering
- ☐ Methylprednisolone or equivalent 500 mg-1000 mg / day pulsed for 3 days **WITHOUT** any tapering
- ☐ Other prednisolone dosages
- ☐ Cyclosporin
- ☐ Cyclophosphamide i.v. bolus
- ☐ Tacrolimus
- ☐ Rituximab
- ☐ I never treat AE-IPF with any immunosuppressive therapy
- ☐ Other immunosuppressive therapies:

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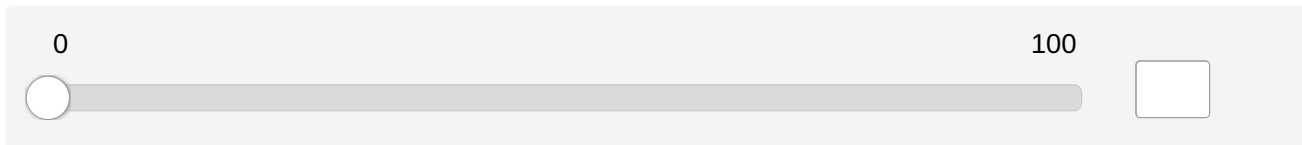
You treat AE-IPF with other prednisolone dosages.

Please, let us know the dosage:

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You treat AE-IPF with corticosteroids. How long would you treat with steroids in weeks?

0 100



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How would you rank your selected therapies?

(1=the therapy which is used most of time; if you have selected only one answer, please label it as 1)

<input type="text"/>	Prednisolone 1 mg / kg / day, followed by slow tapering (over weeks)
<input type="text"/>	Methylprednisolone or equivalent 500 mg-1000 mg / day for 3 days, followed by slow tapering
<input type="text"/>	Methylprednisolone or equivalent 500 mg-1000 mg / day pulsed for 3 days WITHOUT any tapering
<input type="text"/>	Other prednisolone dosages
<input type="text"/>	Cyclosporin
<input type="text"/>	Cyclophosphamide i.v. bolus
<input type="text"/>	Tacrolimus
<input type="text"/>	Rituximab
<input type="text"/>	I never treat AE-IPF with any immunosuppressive therapy
<input type="text"/>	[Insert text from Other]

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

Do you use other therapies for AE-IPF? **Multiple answers possible**

☐ Polymyxin B Hemoperfusion (or similar)

☐ Recombinant Thrombomodulin

☐ Plasmapheresis / plasma exchange

☐ None of the above

☐ Other therapeutical options:

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

Which antimicrobial therapy do you commence regularly in AE-IPF? **Multiple answers possible**

- | | |
|--|---|
| <input type="checkbox"/> Broad-spectrum antibiotics | <input type="checkbox"/> Aciclovir |
| <input type="checkbox"/> Broad-spectrum antibiotics combined with macrolides | <input type="checkbox"/> Ganciclovir |
| <input type="checkbox"/> Antibiotic treatment only when there is a clinical and/or laboratory indication for a bacterial infection | <input type="checkbox"/> Antimycotics |
| <input type="checkbox"/> Usually there is no need for an antibiotic treatment | <input type="checkbox"/> I do not agree with any of the answers |
| <input type="checkbox"/> Other: | |

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You use a broad spectrum antibiotic in AE-IPF.

Which broad spectrum antibiotic is your preferred choice?

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

A patient with pre-diagnosed IPF presents to you with AE-IPF **without** any previous antifibrotic treatment and eligible for such a therapy. What would be your choice as antifibrotic treatment ?

- ☐ I would preferentially initiate Nintedanib
- ☐ I would preferentially initiate Pirfenidone
- ☐ I would initiate either antifibrotic without preference
- ☐ I do not see an indication for antifibrotic therapy at all in this situation

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You chose to initiate a therapy with Nintedanib in the patient with AE-IPF. When would you start the treatment?

- ☐ Immediately
- ☐ Only after stabilization of the patient

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You chose to initiate a therapy with Pirfenidone in the patient with AE-IPF. When would you start the treatment?

- ☐ Immediately
- ☐ Only after stabilization of the patient

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You chose to initiate a therapy with either antifibrotic without preference in the patient with AE-IPF. When would you start the treatment?

I would initiate ...

- ☐ Immediately
- ☐ Only after stabilization of the patient

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A patient on treatment with Pirfenidone/Nintedanib (who tolerated the drug sufficiently) presents to you with AE-IPF. What would you do?

- ☐ Pirfenidone/Nintedanib should be continued unchanged
- ☐ Treatment with any antifibrotic drug should be discontinued
- ☐ Pirfenidone/Nintedanib should be continued at a reduced dose
- ☐ Treatment with the current antifibrotic drug should be stopped and the alternative antifibrotic should be initiated if possible
- ☐ Only in case of pretreatment with pirfenidone, this drug should be discontinued and nintedanib initiated. In case of nintedanib pretreatment I would continue with nintedanib due to the reported effects on time to first acute exacerbation
- ☐ Only in case of pretreatment with nintedanib, this drug should be discontinued and pirfenidone initiated. In case of pirfenidone pretreatment I would continue pirfenidone due to the reported effects on respiratory hospitalizations.

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

If you detect signs of pulmonary hypertension (PH) on clinical investigations (e.g. echo, BNP, clinical signs) during an AE-IPF, how would you proceed? **Multiple answers possible**

- | | |
|---|---|
| <input type="checkbox"/> Start diuretic therapy | <input type="checkbox"/> Start PH specific treatment without a confident diagnosis |
| <input type="checkbox"/> Perform a right heart catheterization | <input type="checkbox"/> Evaluate again after stabilization and possibly perform a right heart catheterization then |
| <input type="checkbox"/> Start PH specific treatment after established PH diagnosis | <input type="checkbox"/> I do not consider PH treatment during or after AE-IPF |

Please name PH drug(s) here (if answer 3 or 4 was chosen)

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Would you initiate or increase the dose of an antacid drug (PPI / H2 blocker) when a patient presents with an AE-IPF?

- ☐ Always initiate or increase antacid drug therapy
- ☐ Always make sure the patient receives antacid therapy but not increase the dose
- ☐ Depending on the presence / intensity of reflux symptoms
- ☐ Never prescribe antacid drug for this situation

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

What kind of ventilatory support would you offer critically ill patients with AE-IPF? **Multiple answers possible**

- ☐ Invasive Ventilation for all IPF patients
- ☐ Invasive ventilation only to patients suitable for lung transplantation (LTX) as a bridge to LTX
- ☐ Invasive ventilation only to patients suitable for lung transplantation (LTX) as a bridge to LTX or very selected other patients
- ☐ ECMO for all IPF patients
- ☐ ECMO only to patients suitable for LTX as a bridge to LTX
- ☐ Consultation with lung transplant center in unlisted, potentially suitable patients
- ☐ High-flow oxygen
- ☐ Non-invasive ventilation
- ☐ None of the above

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

Once AE-IPF is diagnosed, how would you talk to your patient and his caregivers?

- ☐ I give them all information including treatment options and average prognosis during / after AE-IPF
- ☐ I tell them an impression about treatment and prognosis but without telling hard facts
- ☐ I prefer not to tell them anything on the fatal prognosis of an AE-IPF and only some impression on treatments

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

Do you consider palliative care during an AE-IPF?

- | | |
|---|------------------------------|
| <input type="radio"/> Always/Usually | <input type="radio"/> Rarely |
| <input type="radio"/> Only on the initiative of the patients/caregivers | <input type="radio"/> Never |
| <input type="radio"/> Sometimes | |

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

What is your general approach after hospitalization for AE-IPF? **Multiple options possible**

- ☐ Send patients to pulmonary rehabilitation (in- or outpatient)
- ☐ Reschedule patient shortly for a follow up at my center
- ☐ If not contacted earlier during course of AE-IPF send eligible patients for lung transplant evaluation
- ☐ Refer patients not suitable for lung transplant to in- or outpatient palliative or hospice care
- ☐ Send patient home to be seen by their GP shortly for a follow up

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

In your experience, what is the estimated 1-year mortality in patients with AE-IPF?

- ☐ <20%
- ☐ 20-50%
- ☐ 50-80%
- ☐ >80%

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

What is your strategy in the case of a planned surgical procedure in patients with IPF with respect to the risk of acute exacerbations? **Multiple answers possible**

- | | |
|---|---|
| <input type="checkbox"/> Surgical procedures can be performed in the same way in IPF patients as in patients without IPF | <input type="checkbox"/> Elective thoracic surgery should only be performed in suitable patients for the diagnosis of an ILD but not for other indications (e.g. lung cancer) |
| <input type="checkbox"/> Elective thoracic surgery for any reason should not be performed | <input type="checkbox"/> Elective thoracic surgery should only be performed in suitable patients for the diagnosis of an ILD or for lung cancer but not for other indications |
| <input type="checkbox"/> If surgery is necessary, use of low tidal volume and avoidance of hyper-oxygenation to try to prevent injury | <input type="checkbox"/> Any elective surgical procedures should be avoided and only be performed in case of an emergency |
| <input type="checkbox"/> Preferentially use regional anesthesia (over general) when possible | <input type="checkbox"/> I do not agree with any of the statements |

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

What measures do you use to try to prevent AE-IPF? **Multiple answers possible**

- | | |
|---|---|
| <input type="checkbox"/> Antifibrotic therapy | <input type="checkbox"/> Azithromycine long term / maintenance |
| <input type="checkbox"/> Vaccination (influenza, pneumococcal, etc.) | <input type="checkbox"/> Cotrimoxazole long term / maintenance |
| <input type="checkbox"/> Antacids medication (PPI, H2 blockers) in all IPF patients | <input type="checkbox"/> Early screening for PH and treatment with PAH-drugs if PH is diagnosed |
| <input type="checkbox"/> Low dose steroids (<10mg) in all IPF patients | <input type="checkbox"/> Pulmonary rehabilitation or other forms of structured exercise therapy |
| <input type="checkbox"/> Anticoagulants in all IPF patients | <input type="checkbox"/> I do not have a preventive strategy |

☐ Others:

Acute Exacerbations of Idiopathic Pulmonary Fibrosis - Questionnaire

Where do you see the need for improvement in AE-IPF? **Multiple answers possible**

- | | |
|--|---|
| <input type="checkbox"/> Improved collaboration between different ILD specialists in general | <input type="checkbox"/> Increased research and study projects on treatment of AE-IPF |
| <input type="checkbox"/> Improved education and training of physicians | <input type="checkbox"/> Improved multidisciplinary strategies for diagnosing and discussions |
| <input type="checkbox"/> Improved education of patients and caregivers | <input type="checkbox"/> Consensus recommendations concerning the therapy for AE-IPF |
| <input type="checkbox"/> Increased research and study projects on understanding the nature of AE-IPF | <input type="checkbox"/> I do not see any need for improvements |

