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Title: Survival in IPF acute exacerbations: The non-steroid approach

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Body: Background IPF acute exacerbations (IPF-AE) have no proven etiology and although they represent the development of diffuse alveolar damage upon usual interstitial pneumonia, international guidelines recommend high dose steroids. However, the prognosis remains unacceptably dismal. Aims and objectives Assuming that IPF-AE is an ARDS of unknown cause we tested the hypothesis whether ARDS treatment and withdrawing or withholding immunosuppression plus broad spectrum antibiotics influence survival (Papiris et al AJRCCM 2012;185:587-8) Methods We studied patients hospitalized in our clinic from 2007 to 2012 with IPF deterioration. Our protocol consists of immediate cessation of immunosuppression (if any), best supportive care, broad spectrum antibiotics and thorough evaluation to detect reversible causes of respiratory deterioration. All patients were followed up for survival; after discharge none received immunosuppression. We analyzed our data using logistic regression analysis. Results Out of 77 admissions, after excluding other causes of deterioration such as respiratory infection, heart failure, sepsis, pulmonary embolism, pneumothorax, disease progression or progression to the final end, 22 (29%) fulfilled IPF-AE criteria. Among them 50% survived; 3 out of 11 (27%) survived in the group previously receiving

immunosuppression whereas 8 out of 11 (73%) in the group not receiving immunosuppression (p=0.033). The median survival of IPF-AE survivors was 11.5 months. Conclusions Survival of patients treated with the non-steroid approach is better than published data. Previous treatment with immunosuppression is related with increased mortality. No administration of immunosuppression post-discharge is related with prolonged survival.