Title: Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia. 7-year follow-up of a rare clinicopathologic syndrome

Dr. Roger Fei 5244 Falkenstern-Ge Roger-Fei.Falkenstern-Ge@rbk.de MD, Dr. Martin 5245 Kimmich martin.kimmich@klinik-schillerhoehe.de MD, Prof. Dr Godehard 5246 Friedel godehard.friedel@klinik-schillerhoehe.de MD, Prof. Dr Andrea 5247 Tannapfel Andrea.Tannapfel@rub.de MD, Dr. Volker 5248 Neumann Volker.Neumann@rub.de MD and Prof. Dr Martin 5249 Kohlhäufl martin.kohlhaeufl@klinik-schillerhoehe.de MD. 1 Pulmonology, Klinik Schillerhoehe, Center for Pulmonology and Thoracic Surgery, Gerlingen, Baden-Württemberg, Germany, 70839; 2 Thoracic Surgery, Klinik Schillerhoehe, Center for Pulmonology and Thoracic Surgery, Gerlingen, Baden-Württemberg, Germany, 70839 and 3 Pathology, Ruhr University of Bochum BG, Bochum, Nordrhein-Westfalen, Germany, 70839.

Body: Introduction Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH) is a rare clinical pathological syndrome. There have been only 49 cases of DIPNECH reported in the literature so far. Aims and objectives DIPNECH is confined to the airway mucosa without penetration through the basement membrane and appears in a diffuse pattern; little is known about the clinical course and treatment of DIPNECH. Methods DIPNECH is primary proliferations and can combine with obliterative bronchiolitis and bilateral reticulonodular infiltrates. The diagnosis requires the presence of hyperplasia of the airway neuroendocrine cells without invasion beyond the basement membrane. If DIPNECH breaks through the basement membrane and invades locally, this is called tumorlets, whereas nodules causes by DIPNECH > 5 mm in diameter are classified as carcinoid tumors. Results During the 7 years follow up, the patient was repetitively treated with oral steroids because of coughing and dyspnea. The oral steroid therapy was finally replaced by inhaled steroids. All CT- scans over a 7 –year- period showed stable disease without progression of the bilateral pulmonary lesion. Conclusions We present a 7-year-follow up of DIPNECH with stable disease by a non-smoking male patient. The long-term follow up is necessary because there is an association between DIPNECH and peripheral carcinoid tumours. The majority of patients presenting with DIPNECH are middle-aged females with symptoms of cough and dyspnea. In general the clinical course remains stable, however progression to respiratory failure dose occur.