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Somatostatin Analog Use in Patients with Diffuse Idiopathic Pulmonary Neuroendocrine Cell Hyperplasia (DIPNECH)

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BACKGROUND: Diffuse idiopathic pulmonary neuroendocrine cell neoplasia (DIPNECH) is a rare pulmonary condition, characterized by the diffuse proliferation of neuroendocrine cells in the respiratory epithelium. Patients typically present with cough and dyspnea. A diagnosis of DIPNECH is not usually associated with malignant behavior but can be diagnosed in patients with multifocal bronchial neuroendocrine tumors. There are limited data on the management and treatment of DIPNECH; however a few case studies have noted that somatostatin analogs (SSA) may control symptoms.

METHODS: We retrospectively reviewed the records of patients treated at a large referral center evaluated between 1/2008 and 9/2018 with a clinical and/or pathological diagnosis of DIPNECH who received treatment with a SSA.

RESULTS: 26 patients were identified, all of whom were female with radiographic (CT) and pathologic confirmation of disease (69% surgical, 31% biopsy). Median age was 60 years (44-78), 7 (27%) had a history of smoking and all symptomatic at baseline with cough, dyspnea, wheezing, and/or palpitations. Patients presented with a history of symptoms ranging from 6 months – 40 years, with the majority being less than 5 years (38%) at diagnosis. Median duration of SSA (lanreotide LAR or octreotide LAR) was 17 months (1-145 months). 54% of patients experienced no toxicity with SSA, and the 46% who did, experienced diarrhea, cramping and abdominal pain. 2 patients developed gallstones requiring discontinuation of SSA. 38% of patients did not require

additional supportive medications for symptoms; the remainder of patients most frequently also used a steroid inhaler. 21 patients (81%) experienced improvement in their symptoms (10 mildly, 2 moderately, 9 significantly).

CONCLUSION: SSAs can be used in patients with DIPNECH to control clinical symptoms, and result in palliation in the majority of cases. Further research is warranted in this subset of patients.