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## Demystifying DIPNECH: Initial Findings from a New Longitudinal Patient Registry

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### BACKGROUND

Diffuse idiopathic pulmonary neuroendocrine cell hyperplasia (DIPNECH) was first characterized in 1992 with an initial case series describing 6 patients with neuroendocrine cell hyperplasia, with cough and dyspnea symptoms. Since that time, DIPNECH has remained a poorly understood and understudied entity with the largest series reporting 61 patients. Based on the prevalence of DIPNECH at our institution, we believe it may be more common than recognized. We therefore sought to further characterize this syndrome in a new longitudinal registry.

### METHODS

We identified adult patients with confirmed DIPNECH under IRB approval at Vanderbilt-Ingram Cancer Center. Variables including demographics, symptom evolution, pathologic diagnosis, radiographic characteristics, treatment history, and outcomes were collected within a RedCAP database. Data analysis was conducted using R. Exploratory endpoints include correlation of baseline molecular biomarkers with clinical outcomes. Analysis of study objectives will be descriptive and hypothesis generating.

### RESULTS

Sixty-one patients were identified, all female and a majority (77%) never smokers. Median age at symptom onset and diagnosis were 56 (44,64) and 61 (52,67) respectively. Prior to diagnosis, 82% of patients reported chronic cough, 48% dyspnea, 9.8% chest pain, 3.3% hemoptysis, and 1.6% recurrent pneumonia. No symptoms were reported in 6.6% of patients. Patients received many diagnoses before DIPNECH, including GERD (49%), asthma (41%), COPD (8.2%), and ILD (4.9%).

Imaging studies included CT (98%), 68-Ga or 64-Cu DOTATATE PET (61%), fluorodeoxyglucose-18 (FDG) PET (20%), and indium-111 (111-In) pentetreotide (6.6%). CT findings included mosaic attenuation (59%), multiple pulmonary nodules (90%), airway wall thickening (9.8%), and bronchiectasis (3.3%). On DOTATATE PET, 44% demonstrated avid disease, 7.7% mixed, and 49% no avidity.

Concurrent lung neuroendocrine tumors (Lu-NETs) were identified alongside DIPNECH in 70% of patients, with 86% typical carcinoids. More than half (58%) of these patients had multiple identified Lu-NETs.

Post-diagnosis, 48% were treated with somatostatin analogs (SSA), 38% observation alone, and 8.2% systemic steroids. Spirometry at diagnosis was restrictive in 24%, obstructive in 43%, and normal in 32%. After SSA, 71% of patients reported improvement in cough and 70% improvement in dyspnea.

There was no significant difference in post-treatment spirometry findings. Only one patient had died at time of submission, with a minimal median survival time of 8 years.

## CONCLUSIONS

These findings highlight the indolent natural history of DIPNECH and the clinical challenges associated with its diagnosis. Similar to prior series, this registry suggests modest improvement in pulmonary symptoms with SSA treatment. DIPNECH remains a poorly understood diagnosis, and this registry will be expanded with other centers to better characterize disease burden and outcomes.

## ABSTRACT ID 28666