ASO Author Reflections: Utilizing National Databases to Identify Areas of Future Focus When Studying Rare Tumors

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Due to the rarity of some tumors, data are limited to small case series of 10–15 patients seen over a several-decades-long period. As a result, these tumors are often grouped with histologically and/or anatomically similar tumors for staging and prognostic information. One potential solution comes in the form of national databases such as the National Cancer Database (NCDB), which can provide a window into these rare diseases. Our paper used this strength of the NCDB to investigate 872 patients with ampullary neuroendocrine tumors (NETs). Up to this point, the largest series of patients with ampullary NET was published by Schmocker et al. and included 15 patients from The Johns Hopkins Hospital over an 18-year period.¹

The American Joint Committee on Cancer (AJCC) staging guidelines provide guidance for treatment algorithms and prognosis. Currently, duodenal and ampullary NETs are grouped together for staging purposes.² We found that patients with duodenal NETs had significantly better survival than patients with ampullary NETs. This was true for both the entire population, as well as a population of patients treated with surgical resection. Furthermore, when stratified by stage, patients with duodenal NETs still had a significantly better survival than patients with ampullary NETs. Ampullary NETs were more likely to have a higher grade, positive lymph nodes, and be larger than duodenal NETs at the time of diagnosis.³ These data suggest that ampullary NETs have a more aggressive

biology than duodenal NETs and subsequently raise concerns about the AJCC staging guidelines. Given the significant discrepancy in prognosis, it would be preferable to stage ampullary and duodenal NETs separately.

With the development of national databases, these rare tumors have become easier to study. The granularity is limited by the confines of the database, but it can be used to guide future, in-depth clinical and translational research with sequencing and bioinformatics. As a community, we should strive to collect and store tissue from every operation so that when database studies identify an area of investigation, we are able to move forward with the research. Collaborations between cancer centers and participation in national tissue banks is an easy first step. By characterizing these rare tumors on a genomic and molecular level, we can better inform treatment strategies and provide more specific prognostic data to patients.

DISCLOSURES Samantha M. Ruff and Danielle K. DePeralta declare no conflicts of interest.

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