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MIBG Avidity and Progression-Free Survival in Patients with Metastatic Pheochromocytoma are not Dependent on Germline SDHx Mutation Status

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BACKGROUND: Treatment options for patients with metastatic pheochromocytoma/paraganglioma (PCC/PGL) are limited and none are curative. Very little is known about predictors of response to systemic treatment options. Our objective was to identify predictors of response to 131-I-MIBG therapy.

METHODS: We performed a retrospective review of 71 consecutive patients with metastatic PCC/PGL seen in a single center between January 2000 and August 2016.

RESULTS: Fifty-five patients had 123-I-MIBG scans and 45 were positive. Interestingly, there was no difference in MIBG avidity based on primary tumor location (p=0.175) or between patients with <i>SDHx<i/> mutation (N=28; 26 SDHB, 1 SDHD, 1 SDHA) compared to those without SDHx mutation (N=22; 1 NF1, 21 with no mutation identified) (p=0.732). Of the 45 patients with avid disease, 51% were female (n=23) and 84% (n=38) were treated with 131-I-MIBG. The mean age at treatment was 50.9 years. The mean time from initial diagnosis of PCC/PGL to metastatic disease was 5.8 years (range 0-24.5) and did not differ between those with SDHx mutation (n=20; 19 SDHB, 1 SDHD) and those without (n=12;

1 NF1, 11 with no mutation identified) (5.3 vs 6.3 years; p=0.683). The median clinical progression-free survival (PFS) was 34.8 months (95%CI 12.3-58.3). There was no difference in clinical PFS based on SDHx mutation status, primary tumor location or high vs low dose treatment (p=0.589, p=0.211, p=0.463, respectively). Limitations of this retrospective study include small sample size and lack of formal RECIST criteria.

CONCLUSION: These data are interesting as the results demonstrate no clinical predictors of response to MIBG therapy and do not support the notion that SDHB mutations carriers with metastatic PCC/PGL are less likely to be MIBG avid and have a decreased response to MIBG therapy. In summary, these data suggest that all patients with MIBG avid metastatic PCC/PGL may benefit from MIBG therapy.