

antimetatypantibody is added and incubated as 2nd reaction following a wash. Then substrate solution is added after washing immunocomplex. The resulting reaction signals are proportional to the amount of aldosterone in the sample allowing quantitative determination of in serum or plasma sample. The overall reaction is completed within 30 min. **Results** Limit of blank (LoB), limit of detection (LoD) and limit of quantitation (LoQ) of our NC-CLEIA aldosterone assay were 0.09 ng/dL, 0.21 ng/dL and 0.57 ng/dL, respectively. NC-CLEIA aldosterone measurements were linearly well correlated with LC/MS aldosterone measurements ($N = 130$, $y = 1.027x - 0.23$ ng/dL, Spearman's $\rho = 0.996$, $P < 0.0001$). Bland-Altman plot analysis between NC-CLEIA and LC-MS/MS of aldosterone revealed a bias of 0.40 ng/dL with the limits of agreement of -4.60 and 5.41 ng/dL with 95% confidence interval. **Conclusion** Our novel NC-CLEIA aldosterone assay was well-correlated and had only a very low bias with LC-MS/MS method and also was able to accurately quantify low level samples even in essential hypertension patients. This aldosterone assay can be a most equivalent to LC-MS/MS measurement with a low cost of 12 \$ and a short measuring time of 30 minutes.

Pediatric Endocrinology

PEDIATRIC OBESITY, THYROID, AND CANCER

Sporadic MTC in Children: Characterization of a Rare Disease

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INTRODUCTION: Medullary thyroid carcinoma (MTC) is rare in children and is hereditary (hMTC), caused by germline mutations in the *RET* proto-oncogene, in about 95% of cases. Very little is known about sporadic MTC (sMTC) when diagnosed in children/young adults. Our aim was to study the clinical presentation and long-term outcomes of a large cohort of sMTC seen at a tertiary cancer center and to compare sMTC with hMTC in young patients (pts).

METHODS: Through a review of institutional databases, we identified pts diagnosed with MTC \leq age 21 years (y). Charts were retrospectively reviewed and data abstracted. The diagnosis of sMTC vs hMTC was determined based on germline *RET* testing and family history.

RESULTS: We identified 146 pts (53% female), of whom 20 (14%) had sMTC and 126 (86%) had hMTC (80 MEN2a and 46 MEN2b), with a median follow-up of 10 y. (range: 0.08-58, IQR 4.8-18). In pts with sMTC, the stage at diagnosis was I-II in 3/15 (20%) and stage III-IV in 12/15 (80%). Somatic mutations were identified in 11/12 tumors tested (6 *RET* p.M918T, 1 *RET* p.G691S, 2 *RET* deletions p.L629_L633del and p.E632_L633del, 1 *RET* c.2698_2710delinsC, and 1 *CCDC6-ALK* fusion). In contrast to hMTC, pts with sMTC were diagnosed at an older age [mean 18.0 y. \pm 3.4 (range: 10-21) vs 12.9 y. \pm 5.4 (range: 1.5-21), $p < 0.001$], had higher calcitonin

[median 889 (IQR 528-2634) vs 16 (IQR 3-117) x Upper Limit of Normal, $p < 0.001$] and CEA levels [median 186 (IQR 46-468) vs 11 (IQR 4-16) x Upper Limit of Normal, $p < 0.001$], larger tumors [median 2.5 cm (IQR 2-3.7) vs 0.8 cm (IQR 0.4-1.9), $p < 0.001$], and were more likely to be stage IV at diagnosis [73% vs 28%, $p < 0.001$]. sMTC pts were less likely to have bilateral tumors [27% vs 81%, $p < 0.001$] and, at last follow-up, had more persistent structural disease [79% vs 46%, $p = 0.007$] and distant metastases [74% vs 37%, $p = 0.005$]. Death from MTC occurred in 15% of pts with sMTC vs 6% pts with hMTC; median overall survival was not significantly different [30.6 y. in sMTC vs 39.3 y. in hMTC].

CONCLUSION: In this largest reported series of MTC in children/young adults, and the only study to look at sMTC in this population, we identified sMTC in 14% of MTC cases, a higher prevalence than is traditionally recognized but one that is possibly confounded by a referral bias. Somatic mutations were identified in 92% of samples tested, allowing for targeted therapy in those with distant metastases if needed. Compared with hMTC, patients with sMTC presented at an older age with higher tumor markers, larger tumors, and more unilateral disease. At last follow-up, persistent structural disease and distant metastases were more common in sMTC. The differences in clinical presentation and long-term outcomes likely reflect a variable path to MTC diagnosis. In conclusion, sMTC in pts \leq age 21 y. presents at an older age with more advanced disease, frequently has an actionable driver mutation, and may be more common than previously thought.

Pediatric Endocrinology

PEDIATRIC GROWTH AND ADRENAL DISORDERS

Maintenance of Favorable Treatment Effect of Once-Weekly TransCon hGH for Children With Growth Hormone Deficiency: Interim Analysis From the Enlighten Long-Term Extension Trial

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Background Once-weekly TransCon hGH is an investigational long-acting prodrug for growth hormone deficiency (GHD) that consists of 3 components: unmodified growth hormone (hGH; somatotropin), an inert carrier that protects it, and a linker that temporarily binds the two. In the