

No difference was found between TSH levels in the follow-up of the patients with or without a thyroid nodule. **Conclusion:** Thyroid nodule frequency on at HT background was not found to be 13% and the thyroid malignancy frequency 0.67% in our study. A diagnosis of thyroid cancer was made in %5.1 of the patient.

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The Diagnostic, Treatment and Follow-Up Features of Childhood Thyroid Malignancies – A Preliminary Report

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Background: Thyroid cancer is a very rare malignancy of childhood. Approximately they account for 1.5% of all cancers before 15 years of age. In our country, this rate is %0.4 before 20 years of age. **Aims and objectives:** To analyses the clinical features and treatment results of children with thyroid malignancy in Turkey. **Methods:** In this multicentric and retrospective study the demographic and clinical characteristics of 124 children being followed-up for thyroid malignancies from 18 centers between 1991 and 2015 were recorded and analysed. **Results:** The age at diagnosis was 12.9 ± 4.3 years and female/male ratio was 84/40 = 2.1. There was a family history of thyroid disease in 41 cases and thyroid cancer in nine cases. In the first application, cervical mass in 63 cases, thyroid nodule in 34 cases, and both nodule and mass in 14 cases were found. The results of cases that underwent to fine needle aspiration biopsy were benign in seven cases, suspected follicular neoplasm in 17 cases, suspected malignancy in 32 cases, and certain malignancy in 36 cases. Fourteen (11.3%) cases with

disease had distant organ metastasis (13 with lung metastasis). Total thyroidectomy in 110 cases, near total in five cases, central compartment dissection in 20 cases, lobectomy in eight cases and total neck dissection in 18 cases were performed. Pathological examination revealed papillary carcinoma in 94 cases, follicular carcinoma in 14 cases, poorly differentiated carcinoma in two cases, medullary carcinoma in nine cases. Radioactive iodine ablation therapy was applied as low dose in 31% and high dose in 63% of the patients. Recurrence was observed in 14 patients. Recurrence was observed in 14 patients and mean event-free and survival times were 3.3 ± 2.3 and 4.07 ± 3.5 years respectively. **Conclusions:** The diagnostic, treatment and follow-up features of Turkish childhood thyroid malignancies were presented in a large multicenter cohort. These results are expected to contribute to evaluation, follow-up and treatment of these patients.

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New Mutation Causing Systemic Pseudohypoaldosteronism

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Background: Pseudohypoaldosteronism (PHA) is a rare heterogeneous syndrome of mineralocorticoid resistance. PHA type 1 (PHA1) is characterized by neonatal salt loss, failure to thrive, dehydration and circulatory shock. It includes two forms: renal (autosomal dominant), due to mutations in mineralocorticoid receptor coding gene NR3C2, and systemic (autosomal recessive), due to mutations in subunit genes of the epithelial sodium channel (ENaC). ENaC is constituted of three subunits, coded by SCNN1A gene located on chromosome 12p13.31 (alpha subunit), SCNN1B and SCNN1G genes on chromosome 16p12.1 (β and γ subunits respectively). Due to the rarity of the disease, no genotype-phenotype correlations have been established. The systemic form usually presents in the neonatal period with salt loss from kidney, colon, sweat and salivary glands and can show pulmonary symptoms, similar to cystic fibrosis. It is a life-long disease without improvement over time, characterized by life-threatening salt-losing crises that require extensive sodium supplementation and potassium-lowering agents. **Case presentation:** We report the case of a 6-months-old girl with systemic form of PHA1, presented in the neonatal period with dehydration, weight loss, feeding difficulties, hyperkalemia, hyponatremia, metabolic acidosis and elevated plasma aldosterone levels. Clinical conditions improved after elevated sodium and bicarbonates supplementation, administration of ion exchange resins and nutrition with milk formula low in protein and electrolytes.