contributory effect on plasma levels. The tumors weighed between 36 to 103 grams and while in cases #1&3 pleomorphic atypical nuclei with capsular and vascular invasion were reported, the patients had no signs or symptoms of relapse so far. Duration of follow up is 4 to 16 years and no case of relapse has been seen.

Conclusion: Pediatric adrenocortical tumors are multi - hormone secreting neoplasms and have a more favorable prognosis in comparison to adults.

1105

THYROID HORMONE RESISTANCE IN AN IRANIAN FAMILY PRESENTING WITH GOITER AND NEUROLOGIC ABNORMALITIES: REPORT OF THE MUTATION

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Resistance to thyroid hormones (RTH), an inherited syndrome of reduced tissue sensitivity to thyroid hormones is a well known although rare disorder and has not been reported in Iran so far.

Most of the reports concerning the genetic characteristics of the disorder belong to western countries and there are limited reports on thyroid hormone gene study on patients with RTH in Middle East region. Since 1986 we follow the first five patients belonging to a single family with RTH in Iran and reported their clinical pictures and also the effects of short- term treatment with graded dose of T3 on pituitary and peripheral tissue markers. Here we report the result of the gene study on this family.

DNA was extracted from peripheral white blood cells of the family in Tehran, Iran and transported to the Thyroid Study Unit at the University of Chicago. Sequencing of the TR beta gene disclosed a point mutation in ligand binding domain with substitution of Aspartic Acid instead of Glycine at position 345.Same mutation has been reported previously in limited number of patients with similar manifestations.

Conclusion:

1, Glycine to Aspartic Acid substitution is associated with a rather sever form of RTH presenting with big goiter and neurologic manifestations,

2, paucity of the reports on RTH in Asian and African countries are due to the lack of attention to this syndrome and we propose that the diagnosis of RTH should be kept in mind in differential diagnosis of patients who are presumed to be thyrotoxic.

1106

HYPEROSMOLAR, HYPERGLYCAEMIC, NON-KETOTIC ACIDOSIS IN A 17 MONTH OLD - A CASE REPORT AND LITERATURE REVIEW

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Background: Hperosmolar, hyperglycaemic, non-ketotic acidosis (HONK) is a rare initial presentation of diabetes in childhood. It is increasingly reported in the literature in older children with type II diabetes but there are few case reports of younger children with insulin dependant diabetes (IDDM). It is associated with higher morbidity and mortality than diabetic keto-acidosis (DKA), probably secondary to cerebral oedema.

Case: We report a case of this disease in a 17 month old boy with Down syndrome who presented to the ward with a decreased level of consciousness following a short history of being generally unwell. Initial investigation revealed a finger prick glucose of “Hi” with a pH of 7.24. A presumptive diagnosis of DKA as first presentation of IDDM was made. However, subsequent investigations revealed a plasma glucose of 95 g/dl, serum osmolality 400 mosm/kg, with no urinary ketones and the patient was diagnosed with HONK. We discuss the difficulties we encountered in avoiding a precipitous fall in his plasma glucose and in managing his hypernatraemia, which peaked at 171 mmol/l.

Conclusion: HONK is a rare but serious first presentation of diabetes in children. Conventional DKA management is likely to result in a precipitous fall in plasma glucose and exacerbate electrolyte disturbances, increasing the risk of cerebral oedema. Current management is based on data extrapolated from adult studies and a few case reports and involves much lower doses of insulin with cautious fluid replacement. Knowledge of this condition should reduce the risk of serious complications.