has been previously reported in the past. Detection of a high rate and variability of ETAs associated with CH necessitates a vigilant clinical and diagnostic approach to all babies with CH.

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SPECIFICITY OF MEDULLARY THYROID CANCER IN SIPPL'S AND GORLIN'S MEN SYNDROMES IN PEDIATRIC PATIENTS (8 CASES)

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Methods 31325 thyroid patients were operated in the St.-Petersburg Center of Endocrine Surgery and Oncology during 1974 – 2015. Among them 18 (0.06%) cases of MEN were detected, including 8 (0.02%) children. The diagnosis of MEN-2 was confirmed by genetic investigation in all 8 children (3 girls, 5 boys) 4–18 years old. Genetic research confirmed syndrome in 7 children, syndrome – in 1 girl 10 years old.

Results and discussion The reason for examination of 6 children was MEN identified in adult family members operated on for medullary carcinoma. In 2 cases genetic study was conducted after detection of medullary cancer in pediatric patients. Three patients operated on in childhood belonged to the same family, 5 members of which had syndrome MEH-2a in three generations. Multifocal medullary thyroid cancer with cervical metastases was detected in a girl with MEN-2b. Before surgery, 4 (50%) children did not have manifestations of medullary cancer. Microcarcinoma identified intraoperatively in 1 boy. Increase of the calcitonin levels and thyroid nodules were diagnosed in 4 cases. All 8 patients underwent thyroidectomy, in 7 cases – with central lymphadenectomy. Lateral lymphadenectomy was performed in 2 cases, including 1 Gorlin's syndrome (RET p.M918T). Histological examination showed normal thyroid in 2 children, C-cell hyperplasia – in 1, medullary cancer – in 5. Multifocal medullary cancer T4N1bM1 was revealed in the patient with MEN-2b. She was operated on 3 times for relapses lymphatic node metastases removing. Medullary thyroid carcinoma and Parathyroid adenoma together were removed in 1 child.

Follow-up results were search in all our pediatric patients operated on during 1989 – 2018 years. All patients are alive. Only in one case we observed hypercalcytoninemia up to 1000 pg/ml after 3 repeated operation on the neck and bayside adrenalectomies.

Conclusion Thyroidectomy had prophylactic character only in 3 children 4–10 years old. Early thyroidectomy (under 5 years old) has prevented the development of medullary thyroid cancer in children and adolescent.

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TRANSIENT NEONATAL HYPERINSULINAEMIC HYPOGLYCAEMIA SECONDARY TO A HNF1A GENE MUTATION

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Introduction Monoallelic pathogenic variants in HNF1A are a well-established cause of maturity-onset diabetes of the young 3 (MODY3). These variants are rarely reported as a cause of neonatal hyperinsulinaemic hypoglycaemia (NHH).

Case Description A term infant was delivered by spontaneous vaginal delivery with a birth weight of 3.53 kg (50th centile). He was born to a Gravida 2 Para 1 mother who had manifested gestational diabetes that necessitated insulin therapy in the first trimester of both of her pregnancies. His older sibling also had transient neonatal hyperinsulinism that fully resolved at 1 month of age.

He was admitted to the NICU for the management of transient tachypnoea of the newborn. The infant developed symptoms of hypoglycaemia at 1 hour of life and blood glucose level was measured at 0.4 mmol/L. He continued to become hypoglycaemic despite high glucose infusion rates (GIR). Critical samples were collected whilst hypoglycaemic (plasma glucose 1.7 mmol/L) on day 3 of life confirming the diagnosis of non-ketotic hyperinsulinaemic hypoglycaemia. Given the family history samples from the infant and his parents were sent to Exeter Clinical Laboratory for hyperinsulinism genetic testing.

He was commenced on diazoxide therapy at 3 mg/kg/day on day 17 of life and following this therapy his blood glucose levels normalised on 4 hourly oral feeds (GIR = 8.3 mg/kg/min). He successfully tolerated an 8 hour safety fast and was discharged home on day 31 of life. At 7 weeks of life he presented unwell, febrile and hyperglycaemic (blood glucose range 18–22 mmol/l). He had a urinary tract infection and was commenced on antibiotic therapy. His diazoxide dose was discontinued. Close blood glucose observation over a 7 day period and a repeat safety fast demonstrated normoglycaemia.

Genetic testing subsequently demonstrated that both the infant and his mother were heterozygous for a pathogenic HNF1A missense variant (NM_000545.6:c.788G>Ap. (Arg263His).

Discussion NHH causes significant morbidities including adverse neurodevelopmental sequelae and mortality if untreated. Transient NHH is commonly observed in infants of mothers who have gestational diabetes. HNF1A gene mutation however is a rare cause of transient NHH. Identification of this genetic diagnosis has had major implications for this infant and his family. His mother is currently being appropriately treated for MODY3 and has been counselled about the 50% recurrence risk in subsequent pregnancies. Both children will be observed closely from late childhood due to their risk of developing MODY3.

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WHEN LESS IS MORE: A CASE OF ADRENAL SUPPRESSION SECONDARY TO TOPICAL CORTICOSTEROID OVERUSE

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Background Atopic dermatitis (Eczema) is a common chronic remittent inflammatory skin condition. Emollients and topical corticosteroids are the mainstay of treatment and are widely prescribed. This report underlines the danger of combining prescribed and homeopathic treatments and the importance of monitoring steroid prescribing.