accepted in clinical practice is practically excluded. At the same time, it is necessary to take into account the anatomical and physiological characteristics of the child's body and the psycho-emotional state of the children.

To assess the severity of the child's condition prior to the development of classic clinical signs, damage qualimetry is recommended, which is essentially a quantitative characteristic of the injury shoginess. The scoring of polytrauma is defined as the sum of points of private injury. The real threat of traumatic shock arises when the severity of damage exceeds 6 points.

The standard for providing therapeutic measures in children with traumatic shock includes: Pain relief; Venous access; Infusion therapy; Transport immobilization; Medical correction; Oxygen therapy.

The system of rendering psychological and psychiatric assistance to children and adolescents allows rendering differentiated psychiatric and psychological assistance, as well as carrying out rehabilitation activities in a more remote period. The training of pediatricians in the field of disaster psychiatry should be considered the most important condition for the further improvement of the entire system of assistance to victims and those affected by emergencies.

P284 A CASE OF FAMILIAL CRANIAL DIABETES INSIPIDUS – IT'S ALL IN THE HISTORY

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Aims Cranial diabetes insipidus is characterised by polyuria and polydipsia secondary to partial or complete deficiency of antidiuretic hormone. Although in most patients non-hereditary causes underlie the disorder, rarer genetic defects in arginine vasopressin (AVP) synthesis have been identified. We describe the case of a 5 year old girl who posed a significant diagnostic challenge, with a suggestive history and strong family history of cranial diabetes insipidus, but non diagnostic biochemistry.

Methods Our patient was reassessed and AVP gene testing was performed.

Results A 5 year old girl, presented with a four year history of polyuria, polydipsia and associated enuresis. Family history was remarkable for suspected familial cranial diabetes insipidus responsive to desmopressin, in the patient's mother, maternal uncle and maternal grandfather. Genetic testing had not previously been undertaken. Our patient had previously been investigated with water deprivation test at age 3 years and was found to have normal biochemistry, without polyuria during the test. She represented at the age of 5 years to our service with ongoing symptoms and underwent repeat water deprivation testing. Serum sodium and osmolality remained normal (max 140 mmol/l and 285 mmol/kg respectively) with water deprivation but she did not concentrate her urine (urine osmolality 222 mmol/kg at the end of water deprivation test). AVP gene testing was requested and identified a heterozygous pathogenic missense mutation c.61T>C, confirming a diagnosis of familial cranial diabetes insipidus. The same mutation was identified in the patient's mother, suggesting autosomal dominant inheritance. She was commenced on desmopressin with excellent response.

Conclusions Genetic testing may be a useful aid in the diagnosis of inherited cranial diabetes insipidus. Since these patients have progressive loss of AVP, they may initially respond normally to water deprivation testing. If the index of suspicion remains high, genetic testing is recommended to guide treatment.

P285 THE ASSOCIATION OF CONGENITAL ANOMALIES IN PATIENTS WITH CONGENITAL HYPOTHYROIDISM IN GOVERNMENT TERTIARY CARE CENTERS UNDER DUBAI HEALTH AUTHORITY, UAE: 2000–2015, A RETROSPECTIVE CROSS – SECTIONAL STUDY

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Background Congenital hypothyroidism (CH) is the most common endocrine abnormality of the new-born with associated extra-thyroidal anomalies (ETAs) been documented in the literature at varying rates. Majority of the patients are diagnosed with CH at an earlier age due to the implementation of the neonatal screening program in many countries of the world. Congenital heart diseases (CHD) are the most frequently reported extra thyroidal anomaly. No data on such associations have been reported from the Middle Eastern region.

Aim To demonstrate the number and describe the variety of congenital extra-thyroidal anomalies associated with congenital hypothyroidism in babies born in UAE.

Methodology The study was designed as a retrospective, cross-sectional study in patients with confirmed diagnosis of congenital hypothyroidism. 204 patients with confirmed congenital hypothyroidism from two tertiary government centres in Dubai under Dubai Health Authority with specialized paediatric endocrinology units were included in the study. Patients with Down syndrome, preterms < 35 weeks gestation and babies with TORCH infections were subsequently excluded from the study.

Results 39% of the included subjects with congenital hypothyroidism had associated extra-thyroidal anomalies. Out of these 25% had single ETA, 12% had multiple ETAs and 2% had associated syndromes. The total sample had a male to female ratio of 1.1:1 and a significant proportion of males from the Arab population of UAE and Middle-east were affected with CH as compared to other ethnic groups ($p = 0.03$). The most common aetiological cause of CH was determined to be dysmorphogenesis (55%) followed by dysgenesis (45%). Amongst the cohort of dysgenesis, males with ectopic lingual thyroid gland were affected more with ETAs as compared to females of the same cohort ($p = 0.02$). The most common ETAs were congenital heart disease (16%) followed by urogenital tract anomalies (14%). A wide spectrum of multiple ETAs was observed as well as a number of syndromes including Sanjad Sakati syndrome, Turner syndrome, William syndrome, VATER association and Congenital Disorder of Glycosylation.

Conclusion The study detected a higher rate of ETAs associated with congenital hypothyroidism in the included population subset with noted differences in the gender, ethnic distribution and etiological types of CH as compared to what

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.