Erythropoietin level in obese children was lower than in children with normal BMI ($17,24 \pm 10.9$ and $36,31 \pm 31,41$; $p < 0,001$), a negative correlation between BMI and erythropoietin level ($r = -0.26$; $p < 0,05$).

Obese children revealed a negative correlation between the level of sVCAM-1 and the level of erythropoietin in the blood serum ($r = 0,48$; $p < 0,05$).

Conclusions Obesity in adolescents characterized by decreased erythropoietin and increased level of endothelial dysfunction markers sVCAM-1 more than 2 times, VEGF-A – more than 12 times compared to adolescents with a normal BMI. Evaluation of the protective role of erythropoietin in the prevention of endothelial dysfunction and its complications is necessary.

208 USE OF GLP-1 ANALOG IN A PATIENT WITH PRADER-WILLI SYNDROME

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Introduction Prader-Willi syndrome (PWS) is a rare genetic disease caused by deletions or imprinting defects in the region 15q11-q13 leading to hypothalamic-pituitary dysfunction, hyperphagia with excessive weight gain and behavioral disorders. Obesity is a hallmark of PWS, with consequently high incidence of impaired glucose tolerance and type 2 diabetes (T2D), particularly after puberty. Liraglutide, glucagon-like peptide 1 (GLP-1) analog is efficient in treatment of T2D, but also exhibits positive effect on body weight reduction and appetite suppression.

Case Report We present a 17-year-old girl with genetically confirmed diagnosis of PWS (46,XX, ish del (15)(q11q13)) who developed TD2 at the age

15 years. She was never treated with growth hormone. Basal-bolus insulin therapy was introduced but despite good treatment adherence the optimal glycemic control was not achieved. She was also steadily gaining weight although efforts were made to limit caloric intake. At the age of 17 years her body weight was 140 kg (+4.66SD), height 157 cm (-1.63SD), BMI 56.8

kg/m² (+4.78SD). Diabetes metabolic control was unsatisfactory (HbA1c 7.7%).

The treatment with liraglutide (Victoza) was introduced at dose 1.2 mg/day with gradual reduction and discontinuation of insulin therapy. Two months later, she lost 3.1 kg and her HbA1c level was 6.1%. She also reported reduced appetite. For the following 1.5 years her metabolic control was excellent (HbA1c below 6.5%), but after reaching nadir of 135.2 kg (BMI

54.85 kg/m²) she started gaining weight again and currently weights 143.2 kg. No side effects of the treatment were noted during follow up.

Conclusion GLP-1 analogs are effective in treatment of T2D in patients with PWS. However, the positive effect on the body weight, BMI and appetite regulation decreased over time. The literature data regarding use of GLP-1 analogs in patients with PWS are scarce, but they all report improved

metabolic control of T2D. Nevertheless, there are conflicting results regarding body weight and BMI improvement. The treatment with GLP-1 analogs seems to be safe and effective option for therapy of T2D in PWS. Further studies are necessary to confirm preliminary results and establish the guidelines for use of GLP-1 agonists among PWS patients.

209 ARE KIDNEY MALFORMATIONS POSSIBLE FEATURE OF MEN2B SYNDROME? – REPORT OF A PATIENT WITH MEN2B, TYPE 1 DIABETES, SITUS VISCERUM INVERSUS AND KIDNEY MALFORMATIONS

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Multiple endocrine neoplasia Type2b (MEN2b) is a rare familial syndrome caused by autosomal dominant mutations in the RET protooncogen. Patients with MEN2b suffer from aggressive form of medullary thyroid cancer (MTC), pheochromocytoma, multiple mucosal neuromas, gangliomatosis of gastrointestinal tract, and a marfanoid habitus, whereas hyperparathyroidism is exceedingly rare.

Aim To present a patient with MEN2b, diabetes mellitus type 1, situs viscerum inversus and hydronephrosis with megacystis-megaureter syndrome and explore possible etiologic associations between those entities.

Case Report Our patient was born from a normal pregnancy, at term. Fetal ultrasound imaging 'in utero' revealed bilateral dilation of ureters (megaureter), hydronephrosis, duplicated ureters, and situs inversus. On the 2nd day of life bilateral percutaneous nephrostomies were inserted. At the age of two years right upper pole heminephrectomy for ectopic ureter and antireflux surgery of the lower ureter were performed. He has been followed-up by pediatric surgeon and nephrologist. Kidney function was normal, he didn't show any symptoms and didn't require any treatment. At the age of 7 years he was diagnosed with type 1 diabetes. When he was 11,5 years old, during regular follow-up visit, an ultrasound examination of the thyroid gland revealed suspicious nodule and brought to attention an unusual appearance of the patient: thick, prominent lips with submucosal nodules, marfanoid body habitus, musculature weakness and hypotrophy, high arched palate- suggesting MEN2b syndrome. Laboratory evaluation (high calcitonin level) and pathohistological examination of extirpated thyroid confirmed metastatic medullary thyroid carcinoma. Molecular genetic analysis found RET-proto-oncogene pathogenic variant: c.2753T>C (p. Met918Thr) confirming MEN2b syndrome. There were no signs of pheochromocytoma.

Conclusion To the best of our knowledge there are no reports on association of MEN2b and type 1 diabetes or reversed position of major visceral organs.

However, there are scarce reports on kidney malformations in patients with MEN2b. Acknowledging a recognised role of RET gene in kidney development, we suggest that kidney malformations might be a feature of MEN2b syndrome that should be looked for.