contain an identical alpha chain leads to stimulation of gonadal FSH receptors by supraphysiological TSH concentrations.

Case Report We present a 8.5-year-old girl referred to endocrinologist due to obesity and growth retardation that have worsened in the past year. Medical history revealed breast growth without pubic and axillary hair at the age of 8 years and menarche at the age of 8.5 years. Physical examination: short stature and generalized obesity (height 110.2 cm, - 3.6 SD; weight 40.3 kg, +1.8 SD), bradycardia (58 bpm), edematous and dry skin, thin hair, hoarse voice, thelarche Tanner stage III and absence of galactorrhea, pubic and axillary hair. The thyroid gland was not palpable. Laboratory tests revealed severe primary hypothyroidism as part of autoimmune thyroiditis (fT4 5.2 nmol/L, ref. 11-19.5; TSH > 100 mIU/L, ref. 0.05-4.8; anti-TPO 250 IU/mL, ref. < 4.3). US: hypoechoic, hypotrophic thyroid without focal changes. Elevated estradiol (72 pmol/L, ref. < 32 pmol/L) and prolactin concentrations (45 ug/L, ref. < 20 ug/L) with normal gonadotropins were detected. Other significant laboratory findings were macrocytic anemia, hypercholesterolemia and elevated CK and LDH concentrations. X-ray of the wrist showed a bone age of 6 years. Pelvic US: enlarged uterus with thickened endometrium (5 mm) and bilaterally enlarged multicystic ovaries.

Conclusion A diagnosis of VWGS was established and L-thyroxine replacement therapy was introduced. It resulted in normalization of laboratory findings, cessation of menstruation and regression of breast glandular tissue. In the following year her height increased for 15 cm and weight was reduced for 10 kg.

223

EXTREME GROWTH RETARDATION CAUSED BY PRIMARY HYPOTHYROIDISM

G Krajnović*, D Mitrović Dittrich, M Stanić, I Krajnović. OB „Dr.Josip Benčević“ Slavonski Brod

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A twelve-year old girl was hospitalized because of extremely short stature. She was born from normal pregnancy and delivery weight was 3000 g and height 51 cm. She was not ill. Her psychomotoric development was normal. At the age of seven she was 115 cm tall (3. ct), and now, at the age of twelve 118.5 cm (Z score -4.64). She did not visit her doctor at that period. She was extremely short, her skin was dry, she was amimic, bradycardial, she had slowed and irregular tooth growth, and she did not have any signs of pubertal development.

In laboratory test: fT4 <0.4 ng/dl, fT3<0.1 pg/ml, TSH>100 uIU/ml, high levels of cholesterol, anemia, high levels of AST, ALT, CK, pleural and pericardiac effusion. Bone maturity matched that of a six-year old. ITT showed low production of growth hormone, and MRI of hypothalamic pituitary region was normal. We started treatment with levothyroxine, 50 ug per day, and vitamin D. After a few months her laboratory tests were normal. In the period of one year she has grown 11.5 cm, she has gained 5 kg, her bone maturity has improved and now matches that of a ten-year old, puberty started, her skin was normalized, her mood

has improved, she is smiling more often and achieves better results at school. Her family is not cooperative so we included Social Welfare. We follow her development continuously

Paediatric Gastroenterology, Hepatology and Nutrition

224

IMPAIRED SLEEP QUALITY IN CHILDREN WITH INFLAMMATORY BOWEL DISEASE PRESENT EVEN IN THE REMISSION PHASE AND ATTRIBUTING TO IMPAIRED HEALTH RELATED QUALITY OF LIFE

Ivana Trivić*, Zrinjka Mišak, Sanja Kolaček, Iva Hojsak. Referral Centre for Paediatric Gastroenterology and Nutrition, Children's Hospital Zagreb

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Children with inflammatory bowel disease (IBD) have significantly lower health related quality of life (HRQoL) compared to healthy controls. HRQoL presents a broad, multidimensional concept compromising one's physical health, psychological state, level of independence, social relationships, personal beliefs and relationship to the environment. Good sleep is essential in maintaining health and quality of life (QoL) and plays a role in regulation of immune and neuroendocrine system. The aim of our study was to evaluate the relationship between sleep quality and HRQoL in children with IBD in remission.

A total of 33 paediatric IBD patients in remission (20 boys) aged 15.6 ± 1.9 years were included in the study (disease type: Crohn's disease (CD), n=16, ulcerative colitis (UC), n=15, inflammatory bowel disease-unclassified (IBD-U), n=2). Sleep quality was assessed using the Pittsburgh Sleep Quality Index (PSQI) questionnaire, whilst HRQoL was assessed using IMPACT III questionnaire. Moreover, patients wore a triaxial accelerometer for five consecutive days for objective PA quantification. Anthropometric data and inflammatory markers' values such as C-reactive protein (CRP), erythrocyte sedimentation rate (ESR) and faecal calprotectin values were recorded.

Prevalence of impaired sleep quality (PSQI>5) was 36.4%, with mean PSQI score 4.64 ± 2.21 . Highest mean scores were recorded in the sleep duration (mean score 1.06 ± 0.99), sleep disturbance (mean score 1.06 ± 0.35) and daytime dysfunction (mean score 1.00 ± 0.79) components of the questionnaire.

Mean IMPACT III score was 146.36 ± 17.24 . On average, patients spent 38 minutes in moderate-to-vigorous physical activity (MVPA), and 198 minutes in light physical activity (LPA) per day. PSQI score negatively correlated with IMPACT III score (coef. -0.446, $p < 0.01$); meaning that the more significantly impaired sleep quality the more impaired QoL; and with time spent in LPA (coef. -0.482, $p < 0.01$). Interestingly, faecal calprotectin only positively correlated with sleep disturbance score (coef. 0.352, $p = 0.048$), but had no significant correlation with the total PSQI score. No correlation was found between anthropometric and other laboratory parameters, MVPA and PSQI and IMPACT III scores.

More than a third of paediatric IBD patients suffer from poor sleep quality even in the remission phase. Further studies

investigating the relationship between sleep quality, HRQoL and possible presence of subclinical inflammation in paediatric patients with IBD are warranted.

225

MODERATE-TO-VIGOROUS PHYSICAL ACTIVITY IS ASSOCIATED WITH INCREASED MINERAL BONE DENSITY IN CHILDREN WITH INFLAMMATORY BOWEL DISEASE

Ivana Trivić*, Sara Sila, Ana Tripalo Batoš, Zrinjka Mišak, Sanja Kolaček, Iva Hojsak. *Referral Centre for Paediatric Gastroenterology and Nutrition, Children's Hospital Zagreb*

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Inflammatory bowel disease (IBD) in children is associated with malnutrition and growth failure. Body composition alterations, such as bone mass deficits, decreased bone mineral density (BMD) and reductions in lean body mass (LBM) have been described. Physical activity (PA) plays an important role in normal growth and development. Moderate-to-vigorous PA (MVPA) has beneficial effects on muscle mass accrual and bone health. Data regarding PA amongst children and adolescents with IBD are scarce. The aim of our study was to evaluate the relationship between PA and body composition in children with IBD in remission.

A total of 33 paediatric IBD patients in remission (20 boys) aged 15.6 ± 1.9 years were included in the study (disease type: Crohn's disease (CD), $n=16$, ulcerative colitis (UC), $n=15$, inflammatory bowel disease-unclassified (IBD-U, $n=2$). Total body less head (TLBH) dual energy X-ray absorptiometry (DXA) was used to measure BMD, expressed as age- and sex-based Z-scores, and to assess fat mass (FM) and LBM, expressed in grams and as age-, sex- and height-based Z-scores. Patients wore a triaxial accelerometer for five consecutive days for objective PA quantification. Daily caloric intake was assessed using a three day food intake record.

Mean BMD Z-score was -0.41 ± 0.88 ; a third of patients had reduced BMD. Mean FM was 15718 ± 6367.2 g; fat mass index (FMI) Z-score -0.40 ± 0.96 ; LBM 37031.2 ± 8596.4 g; mean lean body mass index (LBMI) Z-score -1.83 ± 1.27 . On average, patients spent 38 minutes per day in MVPA. BMD Z-score positively correlated with body height (coef. 0.502, $p=0.003$), body weight-for-age Z-score (coef. 0.742, $p<0.001$), body mass index (BMI) z-score (coef. 0.558, $p=0.001$). BMD Z-score positively correlated LBMI Z-score (coef. 0.758, $p<0.001$) and minutes spent in MVPA (coef. 0.513, $p=0.004$). LBMI Z-score positively correlated with Z-score (coef. 0.758, $p<0.001$ and negatively correlated with cumulative corticosteroid dose (mg/kg/y) (coef. -0.436 , $p=0.011$). No differences regarding anthropometric and body composition parameters were observed between types of IBD. Female patients had statistically lower total LBM compared to male patients, but there was no difference in LBMI Z-scores. No correlation was found between daily caloric intake, daily protein, calcium, phosphorus and vitamin D intake and anthropometric, body composition and bone health parameters.

Positive correlation was found between BMD and lean body mass, as well as between BMD and minutes spent in MVPA. Intervention studies investigating a causal relationship between PA and favourable body composition in paediatric patients with IBD are warranted.

226

METHEMOGLOBINEMIA IN 2 EXCLUSIVELY BREASTFED INFANTS WITH FOOD PROTEIN-INDUCED ENTEROCOLITIS SYNDROME

Antonella Geljić*, Iva Hojsak. *Children's Hospital Zagreb*

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Food protein-induced enterocolitis syndrome (FPIES) is a non-immunoglobulin E (IgE)-mediated gastrointestinal food hypersensitivity of infancy, characterized by repetitive profuse vomiting, often in association with lethargy, pallor and diarrhea. It is most commonly caused by cow's milk protein (CMP) and soy. Breastfeeding is suggested to have a protective role, and FPIES to CMP in exclusively breastfed infants is extremely rare. We report 2 cases of FPIES to CMP in exclusively breastfed infants who both had methemoglobinemia.

The first patient, a 6-week old male exclusively breastfed infant, was transferred to our hospital due to the worsening of enterocolitis syndrome.

He presented with persistent bloody diarrhea up to 14 times per day, followed by vomiting and fever.

The second patient was an exclusively breastfed female infant who presented at the age of 2 months to our emergency department for persistent diarrhea, vomiting and anorexia without fever.

Both patients at admission were severely ill, pale, adynamic, somnolent and dehydrated. Laboratory findings showed metabolic acidosis, methemoglobinemia, anemia and hyponatremia. After laboratory work-up he was placed on antibiotics, but without significant improvement in clinical status and with persistent watery diarrhea. All laboratory work up was negative (radiologic findings, all cultures (stool, blood, cerebrospinal fluid, urine)), all performed metabolic and immunologic tests were negative.

Highest MHgb level was 5.7% in patient 1 and 9.1% in patient 2. Due to severe diarrhea patients were paced on total parenteral nutrition and breastfeeding was stopped for the whole day. After that small quantities of elemental formula were slowly introduced. Symptoms improved and after several days breastfeeding was reintroduced with strict elimination of CM from mothers diet.

Specific IgE and skin prick test for milk was negative in both patients and atopy patch test was performed only in patient 2 and was negative. In both patient CM challenge test was performed at the age of 12 month without reaction; since then both patients tolerate milk.

FPIES in exclusively breastfed infants is extremely rare but should be taken into consideration in cases of unexplained severe enterocolitis. Transient methemoglobinemia may occur in such patients and usually does not require treatment.

227

DYSAUTONOMIA IN CHILDREN WITH IRRITABLE BOWEL SYNDROME AND INFLAMMATORY BOWEL DISEASE

Antonella Geljić*, Anamari Junakovic, Ivana Trivić, Sara Sila, Magdalena Krbot Skoric, Mario Habek, Iva Hojsak. *Children's Hospital Zagreb*

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To evaluate the presence of autonomic nervous system abnormalities (ANS) in children with irritable bowel syndrome (IBS)