revealed prenatally. Girl was born on 27.05.2018. At birth the diagnosis of BE and multiple dysembryogenic stigmas was made. The girl was transferred to the NMRCCH in an urgent order at first 24 hours of life.

On presentation: BE, total epispadia, divergent labia minora and majora, pubic bones diastasis (up to 4.6 cm). There was a defect on anterior abdominal wall wherethrough bladder mucous membrane was prolapsing. Anus was normal. External sex organs were abnormal, child gender couldn't be identified adequately. The uterus was prolapsing due to anxiety at first 24 hours of life. Acrocephaly, frontal bone persistence, fingers and toes complete syndactyly, uranostaphyloschisis and choanal atresia came under notice.

Results We have performed primary repair of bladder with local flaps on 5th day. Later we performed rehabilitation measures.

Due to molecular genetic testing we revealed heterozygous mutation *c.755C>G (p.Ser252Trp)* in *FGFR2* gene (Apert syndrome, autosomal dominant disorder).

Long-term outcomes were estimated at six months age. The child is tube fed, puts on weight and develops. Urinary system infections took place twice due to vesicoureteral reflux. Note that bladder voiding and storage functions are adequate. Bladder volume is 60 ml and the interval between urinations is about 1 hour. Following step is to perform neurosurgical treatment. Orthopedic surgery should be performed in older age.

Conclusion Bladder exstrophy treatment in newborns even comorbid with other MCAs is optimal and shall be carried out in referral centres. The choice and validation of surgery method for BE treatment demand personalized approach. Multidisciplinary approach in management of these patients is important in the presence of other MCAs.

GP77 OCULOCUTANEOUS ALBINISM IN A NEONATE

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Background Oculocutaneous albinism (OCA) is a genetically heterogeneous group of disorders affecting melanin biosynthesis. It is always an autosomal recessive inheritance and is characterized by decreased or absent pigmentation of hair, skin, iris and retina. Clinical presentation is typically of an infant who does not appear to see at the expected 6–8 week corrected age, often with nystagmus and pale pigmentation. Clinical findings demonstrate grossly obvious iris attenuation and iris transillumination, refractory errors and photophobia. Investigations include genetic testing, Optical Coherence Tomography (OCT) to identify foveal hypoplasia and electrophysiology to establish any excessive crossing of optic nerve fibres at the optic chiasma.

Method A Caucasian female, born to non-consanguineous parents was referred to Paediatric services at one week of age with concerns regarding her pale hair, eyelashes and diffuse pink reflex from both eyes. On examination she had marked iris transillumination without nystagmus and significant hypermetropic astigmatism for which glasses were prescribed. Her father had iris transillumination and nystagmus but met the visual standards to drive. By eight weeks, the infant had

developed nystagmus and was showing some visual awareness. Developmental examination was age appropriate.

Results At initial presentation the expectation of delayed visual maturation was explained to the parents with reassurance that once vision has matured, OCA is a stable condition. Advice was given regarding protection from solar damage as well as the importance of regular skins checks in view of the possible increased risk of skin cancers. The family history and the phenotypic variability between OCA subtypes prompted gene testing to arrive at a molecular diagnosis.

Conclusion The case highlights the importance of early identification so families are aware of the natural history of OCA as well as the need to protect skin from UV damage. Carrier detection and personalised genetic counselling are possible once the disease causing mutations in the family are identified.

GP78 DILATED CARDIOMYOPATHY WITH ROSS III HEART FAILURE INDUCED BY SEVERE IRON DEFICIENCY ANAEMIA POST LONG TERM COW MILK EXCESS

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Aim To present a 1 year and 6 months old girl and 8.2 kg, admitted in the Emergency Department for extremely pale skin, loss of appetite, intense fatigue, muscular hypotonia, tachycardia, polypnea, grade II systolic murmur, hepatomegaly and Ross III heart failure. From the age of 7 months she started to select the food and finally ate only 1 liter/day cow milk. One day before admittance she was not able to wake up from her bed.

Material and Methods The patient performed lab investigations, cardiopulmonary X-ray, cardiac examination and hematology exploration.

Results Cardiomegaly with increased vascular markings was found on X-Ray. ECG revealed tachycardia 148 b/min, alternating with bradycardia, 48 b/min, right atrial dilatation, left ventricle hypertrophy and prolonged QTc 0.45 sec. Echocardiography showed dilated cardiomyopathy, tele diastolic diameter 4.02 cm, EF 0.56, SF 29%, grade III mitral regurgitation, grade I aortic regurgitation, minimal pericardial fluid and slight pulmonary hypertension. Cardiac biomarkers as NT pro-BNP was very high, 8 817 pg/mL, with elevated CK MB 2.51 ng/mL and normal Troponin T. Serology for Parvovirus B19 IgM and Coxsackie was negative, excluding the viral infection as etiology for dilated cardiomyopathy. Severe anemia with 1.9 g/dl Hb was detected, that imposed rapid blood transfusion. The Iron level, ferritin was 0.1 ng/ml, extremely low. Seriated blood transfusions were necessary until Hb achieved 10 mg/dl. Concomitant heart failure treatment was started with Furosemide, Spironolactone, Captopril and low dose of Dobutamine. Iron correction was done with oral products. Folic Acid and Vit C was added in treatment. The evolution was slowly good; gradually normal diet was introduced with meat, eggs, vegetables and fruits.

Conclusions Anemia is cited in the literature to induce dilated cardiomyopathy. Our patient had the most decreased reported value of Hb and Iron, due to long duration of exclusive cow milk daily alimentation. This situation

conducted to severe dilated cardiomyopathy with heart failure. Correction of anemia, iron administration and normal food intake slowly compensated the heart. A cardiology and hematology team was necessary to cooperate in healing this patient.

GP79 IS IT A PORT-WINE STAIN?: VASCULAR BIRTHMARK ON THE FACE POSING A DIAGNOSTIC CHALLENGE

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Infantile haemangioma is a group of vascular tumours arising during infancy which has characteristic clinical and histological findings. Although this type of tumours has a typical presentation, they can mimic the appearance of other vascular lesions.

Two infants, 2 weeks and 5 weeks of age respectively were referred to the dermatology department for 'port-wine stain' as well as 'orbital cellulitis' and also considered as possible 'Sturge Weber Syndrome'. Both infants had vascular lesions in V1 distribution of the trigeminal nerve noticed at birth and in the days prior to presentation developed significant swelling of the affected eyelid. Clinically these are telangiectatic macular lesions present on the eyelid and forehead, extending to the scalp, neck, shoulder and chest in one infant and affecting the perioral area and lower lip with ulceration in the other infant. In both infants there was a significant swelling of the upper eyelid involved resulting in complete closure of the eye. Magnetic Resonance Imaging and Angiography demonstrated intraorbital haemangioma together with abnormalities within the arterial system. One of the infants had involvement of the intracranial arteries and aortic arch qualifying for PHACES syndrome, and the other had involvement of the soft palate. Both patients were commenced on oral propranolol resulting in rapid shrinkage of the eyelid haemangioma, progressive healing of the lip ulceration and regression of the cutaneous component of the haemangioma. Treatment was well tolerated and is expected to continue for 6 to 12 months.

In summary these are two cases of segmental infantile haemangioma with involvement of internal structure mimicking capillary malformation associated with Sturge Weber Syndrome. These cases demonstrate the importance of distinguishing between infantile haemangioma and other vascular lesions to ensure early commencement of appropriate treatment as well as identification and management of internal involvement.

GP80 ONCE MORE ABOUT CUSTOMIZED VERSUS POPULATION-BASED GROWTH CHARTS: HOW TO ASSESS PHYSICAL DEVELOPMENT OF THE INDIGENOUS CHILDREN OF THE NORTH?

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Introduction Despite evidence of race/ethnic differences in growth, the WHO's position nowadays is based on the understanding that all children who were breastfed as infants, grow similarly and a single set of charts can be used to judge growth in any child. The population-based growth charts (PBGC) in assessing physical development of the indigenous children of the North (ICN) lead to ambiguity: pediatricians must recommend correcting their diet because of malnutrition, but to change the diet means to face many associated health issues. The aim of this study is to assess the appropriateness of PBGC for monitoring of growth of ICN.

Methods We compared the dynamics of body weight, height, body mass index and arterial pressure of the Nenets, Khanty, Komi and Slavic children – inhabitants of the Yamal-Nenets Autonomous region (n=5940, age 3 – 17 years) as well as of the Sakha, Slavic and 5 ethnic groups of ICN living in Yakutia (n=278793, age 0 – 17 years). For some of them, the dimensions of inner organs (sonography) and salt taste sensitivity were also analyzed. We used standard methods of parametric statistics.

Results We revealed that in most ICN the body length and mass, being at birth the same or higher than in non-indigenous folks, after the age of 3 years became significantly lower. Both systolic and diastolic arterial pressure in Nenets children become lower than in non-original settlers beginning from the age of 10 years although the arterial pressure in ICN living in Yakutia is higher than in non-original settlers beginning from the pre-school age. There are also differences in salt taste sensitivity (in some groups of ICN lower) as well as in the sonographically determined liver (in ICN bigger) and in the aorta (in ICN wider) sizes.

Conclusions Understanding that racial/ethnic-specific charts are now not recommended because the differences in growth among racial/ethnic groups are shown to be the result of environmental rather than genetic influences, we still must conclude that PBGC may not be optimal for ICN. Customized growth charts adjusted for race/ethnicity are more appropriate. Further research is needed to investigate the benefits and harms of using customized charts for monitoring the growth of ICN. The use of a single standard in ICN is not justified and the claim that 'child populations grow similarly across the world's major regions when their needs for health and care are met' is probably just an assumption.

GP81 EARLY INTERVENTION TO SUPPORT PRETERM INFANT-PARENT INTERACTION AND DEVELOPMENT: RESULTS OF A RANDOMISED CONTROLLED TRIAL ON MATERNAL SENSITIVITY, SOCIAL-EMOTIONAL DEVELOPMENT AND PARENTAL MENTAL HEALTH

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Objective To evaluate the effects of a range of modifying factors on an early attachment focused intervention for parents of very preterm infants in the NICU on maternal sensitivity and infant social-emotional development.