

receiving tube feeds. Mean daily calorie intake on week 1 and week 2 was 24kcal/kg/day and 59kcal/kg/day respectively.

**Conclusions** Reaching nutrition goals is often complicated and delayed in the neonatal pre and post-operative period. This is evident here with delays in provision of nutrition support and in time to reach basic energy requirements. Our results concur with the literature with the greatest decline in WAZ occurring in the neonatal period. During a critical period of growth and development infants with HLHS experience significant challenges and nutritional compromise. It is necessary to develop strategies and guidelines directed at improving the nutritional intake and status of this vulnerable group.

## OC26 FEATURES OF GUT MICROBIOTA COMPOSITION IN OBESE ADOLESCENTS

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**Introduction** Obesity is a multifactorial disease, which may be caused by genetic, psychological causes, obesogenic lifestyle including disturbance in energy balance and a sedentary lifestyle. Gut microbiota takes part in digestion of plant polysaccharides, and also it produces a strong influence on lipid and cholesterol metabolism, therefore gut microbiota dysbiosis may lead to metabolic dysfunction including obesity. Therefore, we made an aim to detect dominant types of gut microbiota at patients with obesity and normal weight and evaluate biochemical parameters of serum alanine transaminase (ALT) and serum alkaline phosphatase (AP), which are obesity-associated metabolic risk factors.

**Methods** The total participant's number was 40. 18 subjects were obese adolescents (BMI=32,57±4,29 kg/m<sup>2</sup>), the 22 subjects were healthy adolescents (BMI=20,01±1,66 kg/m<sup>2</sup>). These two groups were comparable by ethnicity (all participants are Caucasians), gender (10 males and 8 females at the obese group; 13 males and 9 females at the control group), and age (mean age was 14.75±1.52 yrs for obese normal weight and 14.73±1.55 yrs for the normal weight group).

Metagenome sequencing of V3-V4 variable regions of 16S rDNA were done by Novogene Company (China). Concentration of serum ALT and AP were measured using Mindray Automatic Biochemistry Analyzer. Data were analyzed using the bioinformatics services bri-shur.com. For group comparison the *t*-test was used. Statistical significance was accepted at the  $p \leq 0.05$  level.

**Results** Assessment of OTU in two groups of adolescents – healthy and obesity – revealed the minimal values in the obesity group, the maximal – in the healthy, which showed the decrease in gut microbiome diversity in obesity. There was identified significant difference between gut microbiota composition at the obesity and control groups: the *Dorea* genus (phylum *Firmicutes*) dominated at obese patients ( $p=0.05$ ). Otherwise the groups weren't significantly different from each other, but there was a tendency of *Bacteroides*, *Parabacteroides* (the phylum *Bacteroidetes*) and *Slackia*, *Collinsella* (the phylum *Actinobacteria*) prevalence at the obesity group (the  $p \geq 0.05$ ).

Obese adolescents had the mean ALT as 25,34±13,44, and the mean AP was 143,63±83,69, but this is not statistically reliable ( $p > 0,05$ ). The mean ALT of adolescents without obesity was 18,87±13,26, and the mean AP was 165,76±99,01 ( $p > 0,05$ ).

**Conclusion** We report that obese adolescents had higher levels of ALT and lower levels of AP in their serum, and less diverse gut microbiome communities with higher relative abundance of the main bacteria phyla like *Firmicutes*, *Bacteroidetes*, *Actinobacteria*.

## OC27 NATIONAL NEWBORN SCREENING FOR CYSTIC FIBROSIS: GENETIC DATA FROM THE FIRST 6 YEARS

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**Background** Cystic Fibrosis (CF) is the most common life threatening autosomal recessive multisystem disease in the Republic of Ireland (ROI); with a previously quoted incidence of 1 in 1353 and carrier rate of 1 in 19. Screening for CF was incorporated in the National new born screening (NBS) programme in July 2011 in the ROI. The benefit of early diagnosis is well documented. The purpose of the screening is to identify classic CF cases to allow early diagnosis and intervention and improve prognosis. A cut off point of the top 1% Immunoreactive Trypsinogen (IRT) was taken as an indication for 38 mutation panel molecular screening in the ROI National screening programme to maximise identification of affected typical CF cases and to minimise detection of carriers.

**Methods** All neonates from July 2011 to Dec 2017 who had an elevated IRT on NBS were tested with 38 CFTR gene mutation panel and included in this study. Data from clinical and laboratory database were analysed and cross-referenced with patient charts. The Non-NBS database was used to track down cascade relatives.

**Results** In the first 6 and a half years a total of 5,094 newborns (1.16% of total births in the period) were screened for CFTR mutation. During this period, 170 CF affected cases, 22 CFSPID (CF screening positive, Inconclusive diagnosis) and 325 healthy carriers were identified. Phe508del was the most common mutation (75%) followed by Gly551Asp (9.4%). 95 (56%) were homozygous for Phe508del, 17 (10%) were compound heterozygous for Phe508del and Gly551Asp and 22 (13%) were compound heterozygous for Gly551Asp. Hence, 69% of identified as an affected new-borns are eligible to use orphan drugs. There was one missed diagnosis.

**Conclusion** The National new-born screening programme has been successful with only 1 missed diagnosis and less than 50% of carriers identified than predicted. We identified twice the number of affected new-born carrying the Gly551Asp mutation than previously quoted figures from elsewhere in Western Europe. As these are eligible for new orphan drugs this is important for drug reimbursement nationally. The revised incidence of CF in the ROI is less than previous reports (likely due to net immigration). We estimate the new revised incidence to be 1 in 2570 and the carrier frequency is 1 in 25.