

PS-032a **PERFUSION INDEX AND PULSE OXIMETRY SCREENING FOR SEVERE CONGENITAL HEART DISEASE IN NEWBORN INFANTS. RESULTS FROM A COLLABORATIVE ITALIAN STUDY**

F. Schena¹, ¹I Piccioli, ²F Mosca. ¹Neonatal Intensive Care Unit, Fondazione IRCCS Cà Granda Ospedale Maggiore Policlinico, Milan, Italy; ²Department of Clinical Science and Community Health, University of Milan, Milan, Italy

10.1136/archdischild-2014-307384.328

Background Pulse oximetry (POX) is gaining ground as a screening test for severe congenital heart disease (CHDs) but its sensitivity towards aortic coarctation is low. Pulse oximetry-derived perfusion index (PI) has been proposed as a tool to detect critical left heart obstruction but has never been studied prospectively.

Aim To evaluate the efficacy of a neonatal screening combining PI and POX in a large population and to assess the impact of the test in hospitals with different level of care.

Methods Collaborative prospective study in 16 Italian hospitals. Asymptomatic infants who had not received prior cardiac evaluation were tested before discharge (48–72HoL) for pre- and post-ductal SpO₂ and PI. Cut off: SpO₂ 3%, PI

Results 50244 infants were born during the study period (76.7% in tertiary hospitals). 180 CHDs were detected before screening (142 antenatally, 38 clinically). 42169 newborns were screened. 3 CHDs were identified (2 for low SpO₂, 1 coarctation for low PI). 4 cases (2 coarctations) were missed. False positive rate was 0,45% (0,27% for PI). While in tertiary hospitals 95% of CHDs were identified before screening, in 1°-2° level units only 28% were detected clinically and PI-POX screening added a 46% sensitivity to the sole physical examination.

Conclusion Pre-discharge PI-POX screening provides a significant benefit only in 1°-2° level hospitals, where the rate of clinical recognition is low.

PI is capable to identify cases of aortic coarctation that POX misses but needs further evaluation in populations with a higher rate of missed diagnoses.

PS-033 **WITHDRAWN**

Endocrinology and Metabolism

PS-034 **PLASMA COPEPTIN MAY NOT BE A SENSITIVE MARKER OF PERINATAL STRESS IN HEALTHY FULL-TERM GROWTH RESTRICTED FETUSES**

¹DD Briana, ¹T Boutsikou, ¹S Baka, ¹M Boutsikou, ²M Xagorari, ¹D Hassiakos, ²D Gourgio-tis, ¹A Malamitsi-Puchner. ¹Department of Neonatology, Athens University Medical School, Athens, Greece; ²Laboratory of Clinical Biochemistry-Molecular Diagnostics 2nd Department of Pediatrics, Athens University Medical School, Athens, Greece

10.1136/archdischild-2014-307384.329

Background and aims Vasopressin plays a crucial role in the endocrine stress response to a variety of diseases, including insulin resistance and diabetes. Copeptin reliably mirrors vasopressin levels and is considered a marker of acute endogenous stress and insulin resistance. Intrauterine growth restriction (IUGR) due to placental insufficiency is associated with chronic fetal hypoxia, and with a phase of enhanced fetal/early postnatal insulin sensitivity, followed by later insulin resistance.

Methods Plasma copeptin concentrations were determined by ELISA in 50 cord blood samples from well-characterised non-distressed asymmetric IUGR (n = 30) and appropriate-for-gestational-age (AGA, n = 20) singleton full-term pregnancies. Fetuses were classified as IUGR/AGA, based on customised birth-weight standards adjusted for significant determinants of fetal growth. Doppler studies were indicative of placental insufficiency.

Results Fetal copeptin concentrations were similar in IUGR cases and AGA controls. In the AGA group, fetal copeptin concentrations were elevated in cases of vaginal delivery (p = 0.003). No association was recorded between cord blood copeptin concentrations and maternal age, parity, gestational age or fetal gender in both groups.

Conclusions Cord blood copeptin concentrations are probably not affected by IUGR at term, in the absence of fetal distress, possibly due to a balance between copeptin up regulation by chronic fetal stress on the one hand, and copeptin down regulation in the presence of increased insulin sensitivity, on the other; thus, copeptin may not be a sensitive marker of perinatal chronic stress in healthy asymmetric IUGR infants. On the contrary, cord blood copeptin concentrations seem to primarily reflect perinatal stress associated with delivery mode.

PS-035 **AEROBIC TRAINING BUT NO RESISTANCE TRAINING INCREASES THE SIRT3 CONTENT AND REDUCES ADIPOSITY IN SEDENTARY OBESE MALE ADOLESCENTS**

¹K Vargas-Ortiz, ¹L Jiménez-Flores, ²F Díaz, ³P Montañón-Ascencio, ¹V Vazquez-Perez, ⁴A Figueroa, ¹M Macias. ¹Ciencias Médicas, Universidad de Guanajuato, Leon, Mexico; ²Ciencias Aplicadas Al Trabajo, Universidad de Guanajuato, Leon, Mexico; ³Radiología, Hospital General Regional Leon, Leon, Mexico; ⁴Nutrition Food and Exercise Science, The Florida State University, Tallahassee, USA

10.1136/archdischild-2014-307384.330

Introduction Obesity is a risk factor for mitochondrial dysfunction and cardiovascular and metabolic diseases. In a previous research, we demonstrated that 12 weeks of aerobic training (AT) increased the content of SIRT3 protein in skeletal muscle of sedentary obese adolescents. SIRT3 improves mitochondrial function. There are not studies that have compared the effect of AT and resistance training (RT) on SIRT3 content.

Object To compare the effect of AT versus RT on the content of SIRT3 protein in the muscle of sedentary obese adolescents.

Methods Twenty-seven sedentary obese male adolescents (age: 16.7 ± 0.9 years; BMI: 33.7 ± 4.3 kg/m²) completed a 1-month control period prior to be randomised to one of two supervised exercise protocols: AT (3 days/week, 40 min/day, 70–80% peak heart rate (HR_{peak}) or RT (11 exercises, 2 sets/exercise, 12 repetitions/set) for 12 weeks. Biopsies were obtained from the vastus lateralis muscle before and after 12 weeks of training protocol to analyse the content of SIRT3, PGC-1 α and NRF-1 proteins by western blot. Participants were tested for VO_{2peak}, metabolic, and anthropometric variables.

Results AT increased the content of SIRT3, which was associated with improvements in PGC-1 α content (r = 0.7, p < 0.05) and body fat percentage (r = -0.71, p < 0.05). AT improved waist circumference, fat percentage and VO_{2peak}. RT increased arm and thigh muscle area and strength but did not affect SIRT3, PGC-1, and adiposity.

Conclusions Our data suggest that AT was effective for improving the SIRT3 content and adiposity; therefore, AT may decrease the risk of cardiovascular and metabolic diseases.

PS-036 **FACTORS AFFECTING COMPLIANCE WITH ENZYME REPLACEMENT THERAPY WITH IDURSULFASE IN CHILDREN WITH HUNTER SYNDROME: DATA FROM THE HUNTER OUTCOME SURVEY**

¹M Solano, ²I Morin, ³C Lampe. ¹Department of Neuropediatrics, Fundación Cardioinfantil, Bogotá, Colombia; ²Rare Diseases, Shire, Eysins, Switzerland; ³Rare Disease Centre, Dr. Horst Schmidt Kliniken GmbH, Wiesbaden, Germany

10.1136/archdischild-2014-307384.331

Background and aims Manifestations of Hunter syndrome typically become apparent between 2 and 4 years of age; affected children may be treated with enzyme replacement therapy with idursulfase (Shire). This long-term treatment consists of weekly infusions generally administered over 3 h. Patients may sometimes miss scheduled infusions. This analysis investigated the frequency of, and reasons for, missed idursulfase infusions and stopping treatment in children.

Methods This analysis used data from the Hunter Outcome Survey (HOS), a global, observational registry sponsored by Shire that collects real-world clinical information on the natural history of Hunter syndrome and the long-term effectiveness and safety of idursulfase.

Results As of January 2014, data on missed infusions and stopping treatment between HOS entry and last clinical evaluation recorded in HOS/treatment end (median, 35.4 months) were available for 483 children followed prospectively in HOS aged < 12 years at initiation of idursulfase treatment. The mean time from treatment start to last evaluation/treatment end was 47.2 months. In total, 1046 missed infusions were reported in 135/483 children (28.0%). The most common reasons were illness (for 25.5% of missed infusions), holiday/vacation (10.0%) and caregiver/family issues (7.9%). At last evaluation, 31/483 patients (6.4%) had stopped treatment; the most common reason (38.7%) was the patient's/parents' decision.

Conclusions Analysis of HOS data reveals that a variety of factors affect treatment compliance; the most common reason for missing an infusion was illness. However, 72.0% of children receiving idursulfase did not miss a single infusion during this analysis period, and few children stopped treatment.

PS-037 **MATERNAL BARIATRIC SURGERY AFFECTS NEWBORN BODY COMPOSITION**

¹E Malchau Carlsen, ¹B Kanijo Møller, ²KM Renault, ¹D Cortes, ³KF Michaelsen, ⁴O Pryds. ¹Department of Pediatrics, Hvidovre University Hospital University of Copenhagen, Hvidovre, Denmark; ²Department of Obstetrics and Gynaecology, Odense University Hospital University of Southern Denmark, Odense, Denmark; ³Department of Nutrition Exercise and Sports, University of Copenhagen, Hvidovre, Denmark; ⁴Department of Pediatrics, University of Copenhagen, Hvidovre, Denmark

10.1136/archdischild-2014-307384.332

Background and aims Bariatric surgery (BS) is extensively used and one of few lasting ways to treat obesity. Women in child bearing age also undergo BS; BS-offspring has a lower mean birth weight and an increased risk of being small for gestational age compared to non-BS-offspring. The aim of our study was to assess how BS affects newborn body composition and if BS was associated with offspring aberrant fat deposition.

Methods Pregnant women who previous had Roux-en-Y-gastric-bypass were included. Offspring anthropometric measurements

were collected at birth and total and regional newborn body composition was assessed using dual-energy X-ray absorptiometry. The offspring of BS-mothers was compared to offspring of non-BS mothers. Aberrant fat deposition was defined as the percentage of total fat that was placed abdominally. Multiple linear regressions were used to assess the effect of BS.

Results We included 25 BS-offspring and 293 non-BS-offspring for comparison. There was no difference in maternal pre-pregnancy BMI between the groups ($p = 0.16$). BS-offspring had lower birth weight (-311 g, $p = 0.002$), lower fat percentage (-2.6% , $p = 0.002$), lower lean mass (-260 g, $p < 0.001$) and a lower percentage of total fat placed abdominally (-1.6% , $p = 0.024$). The analyses were adjusted for pre-pregnancy obesity, maternal age, parity, gestational weight gain and newborn sex and gestational age.

Conclusion We observed significant differences in body composition between offspring of women with previous BS compared to those without surgery. The BS-offspring had lower birth weight, fat percentage and lean mass. There was no sign of aberrant fat deposition in BS-offspring.

PS-038 **FINAL HEIGHT IN PATIENTS WITH TYPE 1 DIABETES**

¹AL Rodrigues, ²J Extreia, ³D Amaral, ³R Pina, ³C Limbert, ³L Lopes. ¹Serviço de Pediatria, Hospital Divino Espírito Santo, Ponta Delgada, Portugal; ²Serviço de Pediatria, Centro Hospitalar Barreiro Montijo EPE, Lisboa, Portugal; ³Unidade de Endocrinologia, Hospital Dona Estefânia Centro Hospitalar Lisboa Central, Lisboa, Portugal

10.1136/archdischild-2014-307384.333

Background Type 1 Diabetes Mellitus (T1DM) is the most common metabolic disease in children. Growth parameters are important indicators of child's health.

Objective To evaluate final height of patients with T1DM correlating the metabolic control and disease duration with growth and puberty.

Subjects and methods Retrospective analysis of a cohort of adolescents, aged between 15 and 18 years, with T1DM, followed up to final height at a tertiary Hospital clinic. The variables collected were: age, sex, height at diagnosis, final height, parents' height, pubertal height gain, metabolic control during puberty (mean A1cHB). Statistical analysis was performed using SPSS@v20; results are presented as mean \pm SD.

Results Forty six adolescents were included [59% male (M), 41% female (F)]. Mean age at diagnosis was 9.3 ± 3.5 years. Mean A1cHB was 8.15 ± 1.4 . In 26 patients, T1DM was diagnosed before puberty; in these, the age at the onset of puberty was 10.8 ± 1.5 (M) and 9.2 ± 0.6 SD years (F). Height SDS at diagnosis was 0.5 ± 1.5 (M) and 0.35 ± 1.2 (F). Final height was -0.2 ± 1 (M) 0.08 ± 0.9 (F). Target height was -0.29 ± 1.1 (M) -0.02 ± 1 (F). Patients were significantly taller than their parents at diagnosis ($p = 0,03$), and lost height during follow up to final height ($p = 0,004$) yet final height was within target height ($p = 0,3$). There was no correlation between final height and metabolic control ($p = 0,9$) or duration of diabetes ($p = 0,4$).

Conclusion In spite of a taller stature at diagnosis and variable metabolic control, final height was not compromised, arguing against growth compromise being a major hallmark of deficient metabolic control.