Conclusion: Between cord blood ferritin and studied metabolites.

Results: Principal component analysis (PCA) and partial least squares-discriminate analysis (PLS-DA) showed increased concentrations of amino acids; aspartic, leucine/isoleucine (Leu-Ile) and valine but reduced concentrations of methionine, methio-nine-phenylalanine (Met-Phe) and Phe-Tyr (phenylalanine-tyrosine). They also showed increased acylcarnitines; C4-OH(C3-DC), C0-Carnitine, C2-Carnitine, C5-Carnitine, C5-DC, C18-Carnitine and C16:1. There was no significant difference between IGDM and IPGDM except in C6-carnitine which was significantly lower in IPGDM. IDMs have significantly lower cord blood ferritin than controls (p < 0.001). Cord blood ferritin was negatively correlated with maternal HbA1C (r = -0.314; p = 0.026), maternal body mass index (r = -0.452; p = 0.001) and birth weight (r = -0.42; p = 0.002). There was no significant correlation between cord blood ferritin and studied metabolites.

Conclusion: We report that IDMs have alterations in amino acid concentrations and carnitine shuttle at birth and have low intrateric iron stores. Low intrateric iron stores in IDMs is not related to amino acid and acylcarnitine concentrations at birth.

Assessment of small airway impairment in relation to pediatric asthma control and bronchial hyper-responsiveness

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Bronchial asthma is a chronic inflammatory airway disease that affects the whole airway from central to peripheral. Peripheral airway dysfunction and inflammation could lead to failure of asthma control. The current study aimed at evaluation of small airway function in asthmatic children and its relation asthma control and bronchial hyper-responsiveness. It enrolled 60 asthmatics and 30 controls of comparable age and sex. Small airway impairment (SAI) was assessed using impulse oscillometry (IOS) while bronchial hyper-responsiveness was evaluated using spirometry and IOS pre and post bronchodilator. FEV1 was used as a parameter of disease control and showed that 38.3% were well controlled, 53.4% were partially controlled, and 8.3% were uncontrolled. Mean values of R5, R5-R20, and AX were significantly higher in studied asthmatics before bronchodilator administration compared to controls signifying small airway resistance and higher reactance and they were significantly reduced after bronchodilator administration. SAI was detected in 16.7% of enrolled asthmatics using MEF25/75 < 60% compared to 11.7% diagnosed using IOS parameters and asthma poor control was significantly more prevalent among those asthmatic children compared to those without SAI. HRCT showed abnormal airways in the form of branching and mosaic appearance in studied asthmatics with proven SAI.

Conclusion: IOS is recommended to be used as a complementary tool to spirometry in assessment of pulmonary functions in asthmatic children as a dependable indicator of SAI in such children because of its easier performance and reliable Results.

Expression of the antiapoptotic serum survivin in systemic onset juvenile idiopathic arthritis as an indicator of disease activity and predictor of macrophage activation

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Background: Systemic Juvenile Idiopathic Arthritis (SJIA) is a peculiar auto-inflammatory rather than an autoimmune disease with a clear pathophysiological and clinical differences compared to other JIA subtypes. Macrophage activation syndrome (MAS) is an potentially life-threatening complication that seems to be particularly reported with SJIA. MAS is characterized by an overwhelming inflammatory process driven by excessive expansion of T cells and hemophagocytic macrophages. Failure of the apoptotic pathways is one of the implicated theories in the uncontrolled spread of the destructive joint inflammation in JIA and in the development of MAS. Survivin, an antiapoptotic protein, is involved in the regulation of the cytokines and cell cycle progression. High survivin levels are associated with a significant tissue damage, hyperplastic growth and poor response to treatment.

Objective: Studying the value of monitoring the serum level of survivin as a potential predictive and /or prognostic parameter related to the disease activity, degree of joint destruction and evolution of secondary MAS in patients with SJIA.

Methods: Two groups were enrolled in the study; Group I included 22 previously diagnosed SJIA patients (ACR and International League of Associations for Rheumatology criteria); 12 in remission, 8 in relapse and 2 Diagnosed as MAS and Group II included 20 healthy sex and age matched children serving as a control group. This study was conducted over a one year period of clinical and laboratory follow-up of SJIA patients (Group I). Simplified Disease Activity Index (SDAI) score was used to assess JIA disease activity. Secondary MAS was diagnosed according to ACR, EULAR and Pediatric Rheumatology International Trials Organization (PRINTO) diagnostic guidelines. Enzyme-linked immunosorbent assay (ELISA) was used to assess serum survivin at the onset of the study enrollment and was repeated in case of disease activity or development of secondary MAS. Assessment was time scheduled every 2 months or earlier if a disease activity had evolved.

Results: Throughout the study, Ten Patients (45.45%) suffered systemic and articular activity including MAS patients, one patient had only systemic activity, 5 patients (22.7%) had only articular activity and six patients (27.27%) were in complete remission. Serum survivin, ferritin, ESR and Ferritin/ESR ratio showed higher levels during activity than during remission, and a significantly higher levels in MAS group. Ferritin/ESR ratio above three had a 100% sensitivity and 83% specificity for the diagnosis of MAS (AUC = 0.96) Serum Survivin level above 25 pg/ml had 100% sensitivity and 90% specificity in detection of disease activity (AUC = 0.96). and a serum level above 67 pg/ml had 100% sensitivity and 94.7% specificity in the diagnosis of MAS (AUC = 0.99).

Conclusion: Survivin level is an excellent predictive and prognostic marker showing a significant increment at the time of activity and more significant with the development of secondary MAS.