Nutritional assessment and nutritional rehabilitation in children with bronchiectasis and childhood interstitial lung diseases (ChILD): effects on pulmonary functions and clinical severity

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Background: Nutrition is recognized as modifiable contributors to bronchiectasis and interstitial lung diseases (ChILD) development and progression. Nutritional interventions harness great potential in reducing respiratory illness related morbidity and mortality in the developing world. Aim of the study: This study was done to assess nutritional state of children with bronchiectasis and interstitial lung diseases and to study the effect of nutritional intervention program on their growth, pulmonary functions & on clinical severity of lung disease.

Methods: This is a case control, clinical interventional study included 17 patients with bronchiectasis &13 patients with interstitial lung diseases and 40 healthy children served as controls. Nutritional intervention program for 9 months was done, malnourished patients were given 150% of energy requirements for age- matched healthy children and adolescents by addition of high caloric supplements 1.5 Kcal/ml to well-balanced diet (50% CHO & 20% Protein & 30% fats). Nutritional assessment by Subjective Global Nutritional Assessment, Nutritional dietary history obtained by 24 hours recall, complete anthropometric measurements including weight, height, BMI, and mid arm circumference, body composition using bioelectric impedance analysis, and spirometric pulmonary function testing were done to all patients pre and post nutritional intervention.

Results: 56.67% of studied patients were moderately malnourished and 23.33% were severely malnourished. 66.7% of studied patients were underweight and 50% of patients had stunted growth. All anthropometric indices WAZ, HAZ, BMIZ, z-TSF and z-MUAC of studied patients were significantly lower than control groups (P-value < 0.001). FVC% was positively correlated to BMI (P = 0.045/R = 0.465). Patients had lower body fat % compared to controls (P-value = 0.002). Nutritional rehabilitation significantly improved patient anthropometry, body composition and respiratory symptoms. It also decreased SABA use, number of days of school absence, acute exacerbation attacks and hospitalization. However no significant changes in spirometric pulmonary function tests (FEV1%, FVC%, FEV1/FVC and MEF %) (P-value > 0.05).

Conclusion: Patients with bronchiectasis and interstitial lung diseases (ChILD) showed signs of malnutrition and body composition changes that improved significantly after nutritional intervention program for 9 months with significant improvement in frequency of acute exacerbations and hospitalization.

Bone remodeling in beta thalassemia patients, does it differ between Thalassemia major and intermedia?

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Background: The management of patients with thalassemia has improved markedly over the past few decades with the use of optimized transfusion programs and chelating therapy. With prolongation in life expectancy, it has been observed that this hemoglobinopathy is associated with a variety of bone disorders like deformities, bone pains, growth failure, pathologic fractures, osteopenia, and osteoporosis. High-dose iron chelating therapy may also contribute to osteopenia and osteoporosis. Osteoporosis is a significant cause of morbidity in these patients. It is characterized by low bone mass and disruption of bone architecture, resulting in reduced bone strength and increased risk of fractures. The amino-terminal pro-peptide of type I procollagen (P1NP) is a recently introduced biochemical turnover marker (BTM) that is considered the most sensitive index of bone formation in patients with bone disease of varying origins. We assessed the level of P1NP and bone mineral density as measured by dual X-ray absorptiometry (DEXA) in β-thalassemic pediatric patients for early detection of signs of bone remodeling and assess their correlation to the efficacy of therapeutic interventions (blood transfusion & chelation therapy).

Methods: Our study included 60 thalassemic children and adolescents, regularly following up at the Pediatric Hematology clinic of the Pediatric Hospital, Ain Shams University, 40 of them with thalassemia major and 20 thalassemia intermedia. Their ages ranged between 12 to 18 years and they were compared to 30 age and sex matched healthy controls. All children were subjected to full history taking, full clinical examination, laboratory investigation (CBC, serum Ca, Ph, Alkaline phosphatase and serum P1NP level) and DEXA scanning.

Results: There was significantly lower serum Ca level among TM and TI patients compared to control (P<0.001) with similar serum Ph and Alkaline phosphatase levels. P1NP level was significantly higher among TM and TI patients (9.19 ± 3.94 and 6.30 ± 2.47µg/L, respectively) compared to control group (1.80 ± 1.17µg/L, P<0.001). Bone mineral density was significantly lower among cases compared to control (P<0.001). Bone mineral density of lumber spine and P1NP were not significantly correlated with the type of chelation therapy among cases nor with serum ferritin level. There was highly significant correlation between P1NP and BMD of both groups. ROC curve analysis showed that the cut off value of P1NP level between thalassemia major cases and control was at (1.00), with sensitivity (97.5%) and specificity (100%) and between thalassemia intermedia cases and control, the cut off value was (0.9) with sensitivity of (90.0%) and specificity of (96.67%).

Conclusion: This study highlighted the importance of P1NP for diagnosing bone remodeling and osteopenia among thalassemia patients.

Amino acid and acylcarnitine concentrations in full-term infants of diabetic mothers and their relations to in utero iron status

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Objective: To estimate cord blood amino acid and acylcarnitine concentrations in term infants of diabetic mothers (IDMs) and to assess their relations to cord blood ferritin level.

Methods: We studied 50 term IDMs (cases) and 25 healthy newborns (controls). Thirty-seven (37) cases were infants of gestational diabetic mothers (IGDM) while 13 cases were infants of pre-gestational diabetic mothers (IPGDM). Amino acid and acylcarnitine concentrations were measured in cord blood dried spot samples from all newborns using liquid chromatography tandem-mass spectrometry (LC-MS/MS). Cord blood ferritin was...
Conclusion: We report that IDMs have alterations in amino acid concentrations between cord blood ferritin and studied metabolites. Low in-utero iron stores in IDMs is not related to reduced concentrations of methionine, methionine-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 in-utero iron stores. Low in-utero iron stores in IDMs is not related to amino acid and acylcarnitine concentrations at birth.

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, EULEAR 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 94.74% 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.

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.