**XI SOLANEP International Congress / XV Cystic Fibrosis Latinamerican Congress / XV Brazilian Congress of Pediatric Pulmonology**

**001 ISOLATION, PRODUCTION AND CULTURE OF PRIMARY EPITHELIAL CELLS FROM PATIENT’S AIRWAYS FOR THE TESTING OF NEW DRUGS TO CYSTIC FIBROSIS TREATMENT**

Arthur Henrique Pezzo Kmit (Department of Pediatrics, Center for Pediatrics Research (CIPED), State University of Campinas); Juliana Moreira (Department of Pediatrics, Center for Pediatrics Research (CIPED), State University of Campinas); Adriana Vinogre (Department of Pediatrics, Center for Pediatrics Research (CIPED), State University of Campinas); Carmen Silvia Bertazzo (Department of Medical Genetics, State University of Campinas (Unicamp)); Antonio Fernando Ribeiro (Department of Pediatrics, Center for Pediatrics Research (CIPED), State University of Campinas); Margarida Duarte Amaral (University of Lisbon, Faculty of Sciences, BioISI – Biosystems & Integrative Sciences Institute, Lisboa, Portugal e Genética Médica, Faculdade de Ciências Médicas – Unicamp)

Objective: To isolate and cultivate human primary epithelial cells from airways – Bronchial (HBE), Tracheal (HTE) and Nasal (HNEC) – to drug effect evaluation in Cystic Fibrosis (CF) treatment. Methodology: A. HBEs were isolated from CF patient’s explanted lung and the HTEs were isolated from healthy donor’s expanded trachea fragments, according to the following steps: 1a. It was made a lobe dissection. 2a. Bronchia and trachea fragments were subjected to enzymatic washes containing antibiotics cocktail. 3a. The HBEs and HTEs were scraped from the bronchi and trachea. 4a. Seeded in P-100 plates for cell culturing in BEGM. 5a. After one week the cells were transferred to a plate containing six porous membrane filters. B. HNECs were collected from 3 patients with CF and were performed the following steps: 1b. Nasal brushing in both patient’s nostrils to collect the material. 2b. Placed in appropriate culture medium. 3b. Centrifuged and seeded cells into T25 bottles with irradiated fibroblasts and rho kinase inhibitor. 4b. Cells grown until reaching 60–70% confluency and transferred to filters like step 5a. In filters the cells were cultured for 21 days in an ALI medium for cell differentiation and to form a polarized epithelium to allow the evaluation of the CFTR function. Results: HBEs and HTEs cells growth up to 80% confluence in 1 week and the HNECs took one month to reach the 70% confluence. In filters, at the end of 21 days the HBEs cells seeded on the plate 1 and 2 showed a mean resistance of 1374 ± 185 and 1406 ± 379, respectively. The HTEs cells seeded on the plate 3 and 4 demonstrated a mean resistance of 1581 ± 46 and 1570 ± 51, respectively. Finally the HNECs of patients 1, 2 and 3 had a mean resistance per plate of 1347 ± 373, 1731 ± 466 and 3924 ± 278, respectively. Conclusion: The technique of isolation and cultivation of primary bronchial, tracheal and nasal cells from CF patients has been successfully performed for the first time in Brazil.

**002 EARLY PULMONARY FUNCTION TEST AFTER CYSTIC FIBROSIS POSITIVE NEONATAL SCREENING**

José Maria Gonçalves Neto; Amnile Medeiros Costa; Patricia Fernandes Barreto Machado; Déborah de Araújo Barroso de Pinho; Tânia Wrobel Folescu; Aline Mota Fleming.

Objective: Determine the pulmonary function of children with early diagnosis of cystic fibrosis. Methods: All patients with cystic fibrosis diagnosis who did pulmonary function test, were included. Analog signals of flow, esophageal pressure, airway pressure were obtained and converted into digital values to calculate pulmonary resistance and compliance. The airway flow was measured by an anatomic mask attached to a pneumotachograph (Fleisch 00) and a high-sensitivity pressure transducer (Validyne MP45). Lung compliance was considered abnormal less than 1.1 mL/cmH2O/kg and pulmonary resistance above 60 cmH2O/sec. Data were analyzed using SPSS. The mean values and standard deviations were shown on descriptive analysis for continuous variables with normal distribution. Continuous variables with abnormal distribution were shown by median values, minimum and maximum. The normality of continuous variables was verified using the Kolmogorov-Smirnov test. We used the t test for independent samples to compare continuous measurements between two groups. The Levene test was used to verify the equality of variances. The Mann-Whitney test was used when the normality was not confirmed. Categorical variables were described by absolute frequencies and percentage. The association between categorical variables was measured by test of Pearson. Results: We studied 38 infants with a mean age of 6.5 months and mean weight of 6 kg and 21 were female. Lung compliance corrected for body weight was modified in 23 infants (60.5%) and pulmonary resistance in 14 (36.8%). Conclusion: Cystic fibrosis patients, even those who are asymptomatic, that were sent earlier to pulmonary function test, showed a high percentage of impairment on pulmonary compliance and resistance.

**003 SLEEP DISTURBANCES SYMPTOMS IN CHILDREN WITH SICKLE CELL DISEASE AND ITS ASSOCIATION WITH ENDOTHELIAL DYSFUNCTION**

Regina Terse-Ramos (Escola Bahiana de Medicina e Saúde Pública); Ana Marice Ladeia – (Curso Pós-Graduação em Medicina e Saúde – Fundação Bahiana para Desenvolvimento das Ciências); Rozana Texeira (Departamento de Pediatria – Faculdade de Medicina da Bahia – UFCB); Luísa Danielle Alves de Souza Santos (Faculdade De Medicina da Bahia – UFCB); Tatiane Anunciação Ferreira (Serviço De Pneumologia Pediátrica – Hospital Universitário Professor Edgar Santos – UFBA).

Aim: To describe the symptoms associated with sleep disorders and to evaluate its relationship with endothelial dysfunction in sickle cell disease (SCD). Material: Patients aged 6–18 years old with confirmed HbSS. Sleep disturbance was evaluated using the Sleep Disturbance Scale for Children (SDS) and daytime Sleepiness Epworth Scale (ESS); and endothelial function by flow-mediated vasodilation (FMV) of the brachial artery. Methods: Cross-sectional study involving case group and healthy comparison group recruited from the SCD outpatient clinic. Means and standard deviations and percentages were calculated; the Mann–Whitney U test and Spearman Correlation were used; p < 0.05 indicated statistical significance. Results: The study involved 59 SCD and 62 healthy patients. Mean(s) age of case and comparison groups were 12.6(3.1) vs. 11.6(3.0); p = 0.10, respectively; most
were males in the SCG group (p = 0.01); non-white children were the major composition in both groups (p = 0.13). The case group vs. comparison exhibited lower percentage of FMV (10.7% ± 5.3 vs. 16.2% ± 8.4; p = 0.001) respectively. The mean(sd) of the overall score of the SDSC scale between the case and comparison groups were 44.7 ± 11.5 vs. 45.6 ± 12.7; p = 0.76; in the specific subscales, the excessive sleepiness disorder (DOES) and the sleep breathing disorder (SBD) prevailed in the SCG group: 7.1(3.2) vs. 9.1(3.2), p = 0.02; and 6.7(3.4) vs. 5.2(2.7), p = 0.04 respectively. When considered pathologic scores, children using hydroxyurea presented statistically significant higher mean(sd) values of SBD versus those who did not use and to healthy children: 12.6(2.4) vs. 10.4(1.8) vs. 10.6(1.5), p = 0.01 respectively. No correlation was found between SDSC subscales and percentage of FMV. Conclusion: SCG group had higher values of DOES and SBD comparing to the healthy group; children with sickle cell disease had lower values of FMV. Patients using hydroxyurea presented higher pathological scores in SBD scale.

004 IL-8 GENE: A POTENTIAL MODIFIER GENE OF CYSTIC FIBROSIS SEVERITY

Larissa Legnarin Furlan (Medical student, Faculdade de Medicina de Río Preto); Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State Univ); José Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertuzzo de Souza (Department of Molecular Biology – Center for Research in Biochemistry and Molecular Biology – FAMERP).

Objective: To investigate the polymorphisms(rs4074, rs2227306 and rs2227307) of the IL-8 gene, with CF clinical severity. Material: Enrolled 164 CF patients from our center. Methodology: Restriction fragments length polymorphism was used to detect the pol genotypes. In the study, they were analyzed 45 CF clinical markers. Statistical analyses: Fishers exact, X2 and Mann-Whitney tests. Alpha = 0.05. Results: Of the 161 CF patients analyzed for rs4073: 47 – AA genotype, 73 – AT, 41 – TT. For the 164 CF patients analyzed for rs2227306: 55 – CC genotype, 70 – CT, 39 – TT. For the 88 CF patients analyzed for rs2227307: 16-GG genotype, GT – 40, 32 – TT. For statistical analyses were considered the models: codominant(wild homozygous vs heterozygous vs polymorphic homozygous); dominant(wild homozygous vs heterozygous); recessive(wild homozygous vs heterozygous vs polymorphic homozygous); over dominant(heterozygous versus wild vs polymorphic homozygous). Qualitative variables included: sex, race (white and black), age (<6 months and >6months), symptom onset(<3 months and >3 months), diagnosis(<3 months and >3 months), presence or absence of pulmonary and digestive symptoms, body mass index(BMI), nasal polyps, diabetes mellitus, osteoporosis, pancreatic insufficiency, meconium ileus and bacteria. Quantitative data included: O2 saturation, clinical scores (Bhalla, Kang, Shwachman-Kaulycki) and spirometry. There was association between the rs4073 and co-dominant, dominant and over-dominant models for Achromobacter xylosidans (p = 0.034, p = 0.013 and p = 0.034 respectively) in CF patients with two CFTR mutations identified; for rs2227307 and onset of symptoms in CF patients with two CFTR mutations and codominant model(p = 0.037) and dominant(p = 0.012), presence of digestive symptoms in dominant model (p = 0.026). Conclusion: Rs4073, rs2227306 and rs2227307 pol in IL-8 gene can be considered potential modulators of CF clinical severity.

005 REFERENCE EQUATIONS AND VALUES FOR IMPULSE OSCILLOMETRY SYSTEM (IOS) IN BRAZILIAN HEALTHY CHILDREN AND ADOLESCENTS

Pediatric Pulmonology

Maíra Sampaio de Assumpção (Universidade Estadual de Campinas (UNICAMP) e Universidade do Estado de Santa Catarina (UDESC)); José Dirceu Ribeiro – Universidade Estadual de Campinas (UNICAMP); Renata Mabu Gonçalves (Universidade do Estado de Santa Catarina – UDESC); Renata Martins (Universidade do Estado de Santa Catarina – UDESC); Tatiana Godoy Bobbio (Universidade do Estado de Santa Catarina – UDESC); Camila Isabel Santos Schivinski (Universidade do Estado de Santa Catarina – UDESC).

Objective: To determine the reference equations and values for Impulse Oscillography System(IOS) in healthy Brazilian children and adolescents. Material: All participants were submitted to evaluation of respiratory mechanics using a Jaeger MasterScreenIOS (ErichJaeger-Germany) following American Thoracic Society standards. Methods: Observational cross-sectional analytical study with healthy students aged six to 14 from educational institutions in the Greater Florianópolis area. Approval by the Ethics Committee of the Santa Catarina State University and consent form were met. Biometric data were evaluated (weight, height, body mass index) as predictors and for sample characterization. Three tests were recorded to respiratory mechanics evaluation, with data acquisition for at least 20 sec. For data normality verification, the Kolmogorov-Smirnov test was applied, and Pearson’s correlation test identified the relationship between the predicted values of height, age, and weight and the oscillometric variables of resistance at five and 20 hertz (R5 and R20), reactance (X5), respiratory impedance at 5 hertz (Z5), resonant frequency (Fres), and reactance area (AX). Models were developed using simple linear regression and multiple analyses. Results: After the recruitment of 864 children, 123 subjects with an average age of 10.0 ± 2.4 years for boys and 9.9 ± 2.4 years for girls (p = 0.94) were considered for final analysis. Correlations were identified between the dependent and predictor oscillometric variables, with height identified as having the greatest predictive power in the equations developed for boys in all oscillometric parameters, with a mean adjusted R2 of 46.51%. Age had greater influence on Fres (adjusted R2 = 40.1%) and AX (adjusted R2 = 48.8%) for girls. Conclusions: Considering the six IOS variables studied, six reference equations were obtained for each sex. Height was the most influential predictor variable for most IOS parameters in the population studied.

006 EVALUATION OF THE ADMINISTRATION TECHNIQUE OF MDI AND INTERMEDIARIES TO PARENTS OF YOUNG CHILDREN HOSPITALIZED FOR ACUTE BRONCHIAL OBSTRUCTION

Carlos Kofman; Natalia Soledad Escobar; Carlos Daniel Kofman; Alejandro Manuel Teper

Hospital de Niños Ricardo Gutierrez, Buenos Aires, Argentina.

There are no studies that have evaluated inhalation technique errors in young children previously treated with inhaled steroids and/or bronchodilators that require hospitalization for severe bronchial obstruction.

Objective: To evaluate aspects of the inhalation technique in parents of young children with recurrent exacerbations hospitalized for severe bronchial obstruction. Methods: descriptive cross-sectional study. A theoretical and practical survey was administered to parents of inpatients (3 months to four years of age, with recurrent bronchial obstruction, with previous use of MDI + spacer for at least one month) with wheezing and hypoxemia. Spacer type and administration steps were recorded. Also, degree of maternal education was assessed. Results: 48 parents of 48 patients were evaluated. An MD instruction of the inhalation technique had been given to 83.3%, and periodic assessment had been done to 25% of patients. Non valved spacers were used by 33.3% and 56.3% used non validated holding chambers. Only 27.1% performed a proper hygiene of the intermediaries. Parents made a mean of 1.8 (±0.8) errors but no parent properly fulfilled all the administration steps. The more common errors were: not waiting for at least 30 sec for the second actuation (79.1%), not placing the child in the correct position (33%) and not placing tightly the
facial mask (25%). An inverse correlation between the parent’s maximum academic degree and the number of errors was observed (-0.39, p = 0.007).
Conclusion: Mistakes in aerosol therapy technique with MDI + spacers are quite frequent and of diverse nature in parents of young children with recurrent bronchial obstruction presenting severe exacerbations. It is likely that these errors constitute a cause of treatment failure. Identification of mistakes would be essential to reduce its incidence.

007
INFLUENCE OF SINGLE NUCLEOTIDE POLYMORPHISMS IN WHEEZING AFTER ACUTE VIRAL BRONCHIOLITIS

Alfonso Eduardo Alvarez; Fernando Augusto de Lima Marson; Juliana Santiago; Carmen Silvia Bertuzzo; Clarice Weis Arns; José Dirceu Ribeiro. 
Universidade Estadual de Campinas (UNICAMP)

Introduction: Patients hospitalized for acute viral bronchiolitis (AVB) often continue to present symptoms after discharge. It is not clear what features may influence the maintenance of symptoms and post-bronchiolitis wheezing. Objectives: To evaluate the influence of Single Nucleotide Polymorphisms (SNP) in outcomes during one-year follow-up of patients hospitalized for AVB. Material: Patients hospitalized for AVB in two winter seasons in three tertiary pediatric hospitals. Methods: All patients with AVB, defined by the need for hospitalization for oxygen therapy, in two winter seasons in three tertiary pediatric hospitals were evaluated for epidemiological variables, underwent nasopharyngeal aspirate for detection of virus by PCR technique and blood collection to identify presence of genetic polymorphisms. SNP were screened by AccuFill TM System for the OpenArray® Real-Time PCR Platform. SNP in the following genes were studied: TLR4, TLR2, TLR9, CCL5, IFNA5, NOS2, JUN and VDR. All patients were followed-up for one year. Results: 85 patients (all < 1 year) were followed-up. Outcomes after three and 12 months of discharge (in percentage) were: wheezing (58.8 and 46.1), use of bronchodilators (56.0 and 47.4), use of oral steroids (38.8 and 43.4), need to seek the emergency room (38.1 and 35.1), need of hospitalization (8.3 and 6.4), pneumonia (10.6 and 3.8), 36.5% and 11.8% of patients used inhaled steroids and montelukast, respectively. SNP associated with worse outcome were: rs1927911 (TLR4), rs7656411 (TLR2), rs352162 (TLR9), rs187084 (TLR9), rs1068026 (NOS2), rs2280785 (CCL5), rs2228570 (VDR) (p < 0.05). Conclusions: Infants with AVB have high rates of symptoms after hospital discharge. Some SNP in TLR4, TLR9, CCL5 and VDR gene are associated with worse outcome demonstrating that genetic features contribute to maintenance of symptoms. These findings may contribute to a better understanding of the mechanism of post-bronchiolitis wheezing.

009
THE PROGNOSTIC ROLE OF INTERFERON LEVELS IN THE SEVERITY OF INFLUENZA IN CHILDREN

Marcelo Comerlato Scotta; Ana Paula Duarte de Souza; Bárbara Nery Porto; Leonardo Araújo Pinto; Renato Telêlbom Stein; Rita Mattielo. Pontifícia Universidade Católica do Rio Grande do Sul

Objective: Predictors for severity of influenza infection are not completely understood; as mucosal interferon is the first line of innate immunity against influenza virus, our aim was to assess levels of interferon in respiratory secretions and need of hospitalization in children with Influenza. Methods: We collected nasal wash from children aged less than five years, presenting with influenza-like illness to the emergency room of Hospital São Lucas da PUCRS between June and September 2014. Children with more than 72 hr from onset of illness were excluded. Presence of influenza virus was analyzed through Reverse Transcription Polymerase Chain Reaction and levels of alfa interferon. Results: Sixth-six patients were screened. Median age was 13 months (interquartile range (IQ) 4–25), 60.7% were Caucasian and 57.4% were male. Sixteen tested positive for Influenza. Levels of interferon alfa in respiratory samples were significantly different comparing three patients who needed hospitalization to 13 who were well enough to be managed as outpatients, with median/IQ 0.210 pg/ml (0.175–0.266) versus 1.215 pg/ml (0.524–3.080), p < 0.01. Patients without influenza, hospitalized or not, had interferon levels similar to those admitted with influenza, 0.184 (0.131–0.291). Conclusions: This is the first study correlating severity and respiratory levels of interferon in children with influenza. Our findings highlight the importance of interferon in the immune response against influenza in humans, since complications could be a result of deficient interferon productions due to host or viral factors. Further, interferon levels may become an important prognostic test in clinical practice due to its high negative predictive value for severity, which could lead to more precise interventions. As patients without influenza have low levels of interferon regardless of severity, interferon probably is not important in response against other viruses.

008
AZITHROMYCIN THERAPY IN INFANTS WITH BRONCHIOLITIS REDUCES RECURRENT WHEEZING 3 MONTHS AFTER HOSPITALIZATION: A RANDOMIZED, PLACEBO-CONTROLLED TRIAL

Victória da Avevedo Silveira; Clarissa A. Roza; Fernanda Luisi; Paulo Mário Pitrez; Renato Telélbom Stein; Leonardo A. Pinto. 
Department of Pediatrics, Centro Infantil, Institute of Biomedical Research, PUCRS; Porto Alegre, Brazil

Objective: to test the hypothesis that azithromycin reduces recurrent wheezing and hospital readmission risks 1, 3 or six months after hospitalization by acute viral bronchiolitis (AB). Material and methods: we performed a randomized, double-blinded, placebo-controlled trial in Southern Brazil, from 2009 to 2011 (MACRO Study). Infants (< 12 months of age) hospitalized with AB were recruited in a tertiary university hospital. Patients were randomized to receive either azithromycin or placebo, administered orally, for 7 days. At enrollment, clinical data were recorded. For the present study, main outcomes were wheezing and hospitalization in a follow up, 1, 3 and six months after the AB. Results: Ninety-one patients were included in the follow up (azithromycin 51 subjects, placebo 40 subjects). Hospital readmission risk was not different between the groups studied at 1, 3 and six months after AB. Recurrent wheezing risk was statistically significantly in infants at three months post AB (P = 0.038, OR = 3.63, CI = 1.37 – 9.63). Conclusions: Azithromycin reduces the recurrent wheezing risk three months after hospitalization. Azithromycin therapy should be considered in severe AB and may provide benefit in the reduction of post bronchiolitis recurrent wheezing.

010
REFERENCE EQUATION FOR DISTANCE WALKED IN SIX-MINUTE WALK TEST IN HEALTHY CHILDREN AND ADOLESCENTS

Evandro da Silva Aquino (Pontifícia Universidade Católica de Minas Gerais – PUC Betim; Instituto de Medicina Tropical – USP; Hospital Infantil João Paulo II – FHEMIG); Francielly Dorvina M. R. de Carmo (PUC Minas-Betim); Veronica Priscilla Cardoso da Silva (PUC Minas – Betim); Cristiane Cerchi Coelho (Hospital Infantil João Paulo II – FHEMIG).

The six minutes walking test has been used worldwide to evaluate adults and children as a functional capacity test, although reference values are a key point for all population. Objective: To establish a reference equation for the six-minute walk test in healthy children and adolescent and to evaluate the agreement of the existing equations. Methods: This was a prospective cross-sectional study of healthy children and adolescents randomly selected with aged six to fourteen. Volunteers from five public schools in Betim were evaluated with the six minutes walk test in accordance with the ATS recommendations and had collected anthropometric data. The SPSS version
S10 Abstract

17.1 were used for the statistical analyses. The Pearson and Spearman correlation was performed and multiples linear regressions model. For qualitative variables were used T student test. In all cases the results were considered significant when P < 0.05. Results: A total of 330 children and 162 boys and 168 girls. The distance walked correlated with the variation of heart rate (Pearson correlation R = 0.413, P < 0.001) and work rate (Spearman correlation R = 0.507, P < 0.001). For qualitative variable, sex interferes with the walked distance (P < 0.05). The multiple regression model resulted in the following equation: Distance (m) = 485.55 − (13.86 * sex) + 0.95 * variation of heart rate (bpm) + 3.62 * work rate (km / kg), male = 0 and female = 1. The proposed model correlated with the model by: Li et al 2007 (ICC 0.48 P = 0.000), Geiger et al 2007 (ICC 0.71 P = 0.000). Conclusion: The equation proposed in our study were influenced by gender, heart rate variation and work rate. This study was supported by research initiation background from Pontificia Universidade Católica de Minas Gerais campus Betoim, FIP 8092/2014.

011 PILOT STUDY OF POSTURAL EVALUATION AND THE INTERFACE WITH FUNCTIONAL PARAMETERS IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS

Adriana Della Zuana; Amanda Matos Chaves da Silva; Daniela Garcia Pizzini; DanielleBernini Pere; ElianaTakahama Sakamoto; Marianna Rodrigues da Cunha.
Instituto da Criança – HCFMUSP

Objective: To describe the pilot study results of postural evaluation and association with functional parameters in cystic fibrosis (CF) patients. Methods: This prospective study collected data during routine visits to the CF outpatient clinic. Anthropometry data included weight, height and body mass index (BMI). The postural evaluation was performed using the postural assessment software (PAS/SAPO), which uses a digitized photograph of the patient, allows measuring the position, length, angle and alignment of the body segments of an individual. The strength of the respiratory muscle function (PImax and PEmax) was measured by a manovacuometer. A six-minute minute walk test (6MWT) was performed to assess the functional exercise capacity and lung function testing performed using LIASON® Real-Time PCR Platform. SNP in the following genes were studied: TLR4, TLR2, TLR9, CCL5, IFNA5, NOS2, JUN and VDR. Results: 181 infants (176 <1 year) admitted to hospital were included; 58% male, median age 3.8 months (14 to 622 days); median birth weight 3102 grams (565–4850 grams); 69.5% caused by Respiratory Syncytial Virus. Evaluated outcomes were length of stay, median 6.5 days, duration of oxygen therapy, median 5 days, necessity (34%) and duration of ICU stay, median 9 days, necessity (21%) and duration of mechanical ventilation, median 8.5 days. Death occurred in 5 patients (2.8%). SNP rs4986790 (TLR4), rs352162 (TLR9), rs187084 (TLR9), rs2107538 (CCL5) and rs2228570 (VDR) were associated with worse outcome (P < 0.05). Conclusions: Some SNP in TLR4, TLR9, CCL5 and VDR gene are associated with worse outcome of patients with SAVB demonstrating that genetic features contribute to the severity of the disease. These findings may contribute to a better understanding of BVA.

013 VITAMIN D LEVELS ASSESSED IN CYSTIC FIBROSIS INFANTS DIAGNOSED THROUGH NEWBORN SCREENING

Marina Simões Oliveira; Maria Angela Gonçalves de Oliveira Ribeiro; Maria de Fátima Correia Pimenta Servidoni; Antonio Fernando Ribeiro; José Dirceu Ribeiro; Adyélia Aparecida Dalbo Contrera Toro. Universidade Estadual de Campinas

Aims: To evaluate vitamin D levels in cystic fibrosis infants diagnosed after Newborn Screening and its association to evolution of lung disease. Subjects and Methods: Infants aged 0 to 2 years 11 months 29 days old followed by the Cystic Fibrosis Outpatient clinic and diagnosed after Newborn Screening were included. Information about sun exposure and use of medication were asked to parents and legal guardians, while clinical history, diagnostic and complementary exams were assessed through medical records. A blood sample was collected to assess serum levels of vitamin D using LIASON® 25 OH Vitamin D TOTAL Assay. The data were assessed through Mann-Whitney Test, Fisher-Freeman-Halton Test and Fisher Test using SPSS Statistics 17.0 (p < 0.05). Results: Twenty-six infants aged 1–34 months old (mean 17.12 ± 10.16; median 18.50) participated in the study, 53.8% of whom were male. The mean gestational age was 38.64 weeks ± 1.65 (median 38.00) and the mean birth weight was 2.916 ± 0.440 (median 2.890). The mean age of first colonization by Pseudomonas aeruginosa was 8.38 ± 5.8 (median 7.0) months old, and 5.59 ± 6.11 (median 3.50) by Staphylococcus aureus. Fifty percent of the patients were vitamin D insufficient. The association of vitamin D levels and seasonality was not statistically significant (p = 0.286). The presence of Pancreatic Insufficiency, in 92.3% patients, showed no association to vitamin D insufficiency, present in 45.8% (p = 0.480) of them. There was also no statistically significant association to sun light exposure, though 23.1% of the patients with vitamin D insufficiency were not exposed to sun light (p = 0.115). Conclusion: Vitamin D levels presented no statistically significant association to Pancreatic Insufficiency, seasonality and sun exposure. This may be explained by the small population studied. Further studies with a larger sample and follow-ups are needed to better elucidate the effects of vitamin D insufficiency in Cystic Fibrosis pathophysiology.

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012 SEVERE ACUTE VIRAL BRONCHIOLITIS: A GENETIC ENTITY

Alfonso Eduardo Alvarez; Fernando Augusto de Lima Marson; Juliana Santiago; Carmen Silvia Bertuzzo; Clarice Weiss Arns; José Dirceu Ribeiro. Universidade Estadual de Campinas (UNICAMP)

Introduction: Most patients with Acute Viral Bronchiolitis (AVB) that requires hospitalization for oxygen therapy don’t have any risk factor for the disease. Researchers have studied if genetic features may be associated with severity of AVB. Objectives: To evaluate the influence of Single Nucleotide Polymorphisms (SNP) in outcomes of infants with severe Acute Viral Bronchiolitis (SAVB). MATERIAL: Patients hospitalized for SAVB in two winter seasons in three tertiary pediatric hospitals. Methods: Cross-sectional, prospective cohort study that included all infants with SAVB, defined by the need for hospitalization for oxygen therapy, seen in three tertiary pediatric hospital in two consecutive winter seasons. Patients were evaluated for epidemiological variables and severity of disease, underwent nasopharyngeal aspirate for detection of virus by PCR technique and blood collection to identify presence of genetic polymorphisms. SNP were screened by AccuFillTM System for the OpenArray® Real-Time PCR Platform. SNP in the following genes were studied: TLR4, TLR2, TLR9, CCL5, IFNA5, NOS2, JUN and VDR. Results: 181 infants (176 <1 year) admitted to hospital were included; 58% male, median age 3.8 months (14 to 622 days); median birth weight 3102 grams (565–4850 grams); 69.5% caused by Respiratory Syncytial Virus. Evaluated outcomes were length of stay, median 6.5 days, duration of oxygen therapy, median 5 days, necessity (34%) and duration of ICU stay, median 9 days, necessity (21%) and duration of mechanical ventilation, median 8.5 days. Death occurred in 5 patients (2.8%). SNP rs4986790 (TLR4), rs352162 (TLR9), rs187084 (TLR9), rs2107538 (CCL5) and rs2228570 (VDR) were associated with worse outcome (p < 0.05). Conclusions: Some SNP in TLR4, TLR9, CCL5 and VDR gene are associated with worse outcome of patients with SAVB demonstrating that genetic features contribute to the severity of the disease. These findings may contribute to a better understanding of BVA.
014
INSPIRATORY MUSCLE STRENGTH AS A PREDICTOR OF MAXIMUM OXYGEN CONSUMPTION IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS

Fernanda Maria Vendrusculo (Pontifícia Universidade Católica do Rio Grande do Sul); João Paulo Heinzenmann-Filho (Pontifícia Universidade Católica do Rio Grande do Sul); Rafaela Borges Soares (Pontifícia Universidade Católica do Rio Grande do Sul); Leonardo Araújo Pinto (Pontifícia Universidade Católica do Rio Grande do Sul); Paulo José Caudaro Marostica (Universidade Federal do Rio Grande do Sul); Márcio Vâncius Fagundes Donadio (Pontifícia Universidade Católica do Rio Grande do Sul)

Objective: To evaluate the influence of lung function, respiratory muscle endurance and strength as predictors of maximum oxygen consumption (VO2max) in children and adolescents with cystic fibrosis (CF). Methods: Cross-sectional study. Patients with CF, aged six to 18 years old and who were followed in the outpatient clinics were included. Spirometry, manovacuometry, inspiratory muscle endurance (IME) and cardiopulmonary exercise tests (CPET) were performed. All results were normalized by reference equations, except the IME. All tests followed national and international guidelines. Also, demographic, anthropometric and clinical information through medical records were collected. The influence of the independent variables (FEV1, FVC, FEF25-75%, MEP, MIP and IME) on VO2max was analyzed using a stepwise multiple linear regression model.

Results: So far, 8 CF patients were included. The mean age was 16.5 ± 1.7, BMI (z-score) of −0.32 ± 1.0 and 50.0% were male. There was a mild impairment of lung function. The ventilatory muscle strength presented values within normal range and the average IME (cmH2O) was 67.1 ± 21.8, with a percentage relative to maximum load of 57.5 ± 15.8. As to the criteria in order to complete CPET, 7 patients reached a plateau, 6 reached the maximum heart rate (HR) expected, and all of them interrupted the test due to exhaustion. The HR increased 81.4 ± 26.2 bpm and there was no desaturation in any patient after exercising. The mean VO2max (mL/kg/min) was 43.9 ± 19.4, the exhaled minute volume 64.2 ± 32.3 (L/min) and the ventilatory equivalent (L/min) for O2 25.0 ± 5.4 and for CO2 24.2 ± 5.4. In the multiple linear regression model, only the IME has proved to be a significant variable in predicting VO2max, measured by CPET, explaining 87.0% of its performance. Conclusion: The partial results indicate that the evaluation of IME can be used as a predictor of maximum oxygen consumption in the cardiopulmonary exercise test in children and adolescents with CF.

015
INFLUENCE OF FUNCTIONAL CAPACITY AND RESPIRATORY MUSCLE STRENGTH ON HOSPITALIZATION, AND ANTIBIOTIC USING OVER FIVE YEARS IN PATIENTS WITH CYSTIC FIBROSIS

João Paulo Heinzenmann Filho (Pontifícia Universidade Católica do Rio Grande do Sul); Fernanda Maria Vendrusculo (Pontifícia Universidade Católica do Rio Grande do Sul); Taila Cristina Piva (Pontifícia Universidade Católica do Rio Grande do Sul); Juliana Severo da Silva (Pontifícia Universidade Católica do Rio Grande do Sul); Paulo José Caudaro Marostica (Universidade Federal do Rio Grande do Sul); Márcio Vâncius Fagundes Donadio (Pontifícia Universidade Católica do Rio Grande do Sul).

Objective: To evaluate the influence of functional capacity and ventilatory muscle strength on hospitalization and the use of antibiotics (ATB) over five years in patients with cystic fibrosis (CF). Methods: This is a prospective cohort study. Patients with CF, aged six to 18 years, followed by five years in an outpatient setting, were included. Patients underwent spirometry tests, manovacuometry and six minute walk test (6MWT) when entering the study and in each subsequent year. Participants, who did not undergo some of the tests during follow-up, were excluded. Anthropometrics, clinical data and information on days of hospitalization and use of antibiotics were collected. The patients were divided according to the median found for the results of manovacuometry and the 6MWT. The Mann Whitney U test and repeated measures ANOVA with Bonferroni post-test were performed. Results: The sample consisted of 26 children and adolescents. The mean age was 10.2 ± 2.8, BMI (z-score) of 0.5 ± 1.3 and 57.7% were male. There was a mild impairment of lung function, with respiratory muscle strength and functional capacity preserved. When evaluating the progress of these outcomes, there was a significant decrease in FEV1 and FVC over the five years. On the other hand, there was a significant increase in MIP, while the MEP and the 6MWT remained stable. Patients with a smaller distance covered than 577.5 meters in year one required more hospitalizations over the five years, compared to individuals performing above 577.5 meters. There was no difference in the use of ATB when separated by the distance in the 6MWT. Similarly, there was no difference in days of hospitalization and ATB use when separated by MEP (<83.5 or >83.5) or MEP (<89.0 or >89.0). Conclusions: CF patients with greater functional capacity required fewer hospitalizations over five years of follow-up. There was no influence of the 6MWT and the ventilatory muscle strength on the use of ATB.

016
SEROPREVALENCE OF BORDETTA PERTUSSIS IN PREGNANT WOMEN AND NEWBORNs

Juan J. Roselló; Alejandro Teper; Fernando Pollak; Romina Libster; Manuel Roa Rivarola; Hospital Universitario Austral, Pilar. Buenos Aires, Argentina.

Objective: Whooping cough is a highly contagious disease caused by Bordetella Pertussis. It has a high rate of morbidity and mortality, especially in infants under six months of age. In Argentina the incidence and mortality are on rise in the past 3 decades. Failure to induce lasting immunity to current vaccines and antigenic divergence between local strains and vaccines aggravate the effect of waning immunity. The aim of the study was to compare the level of antibodies against Bordetella Pertussis in the neonate from vaccinated and unvaccinated pregnant women at the third trimester of gestation. Methodology: We designed an observational, cross-sectional study to determine the concentration of antibodies against Bordetella Pertussis. We measured antibodies in mothers with and without vaccination at the third trimester of gestation and their newborns, measured in cord blood. We enrolled 111 mothers and their babies; 35 without vaccine and 76 with vaccine. Antibodies were measured in mothers at the third term of pregnancy and in cord blood at deliver. Antibodies determinations were made with the ABCAM anti Bordetella Pertussis IgG Human ELISA Kit. Chi2 was used to compare prevalence. Results: Infants of vaccinated mothers presented negative Pertussis IgG antibodies in 8% (6/76), while infants of unvaccinated mothers were negative for Pertussis IgG antibodies in 100% (35/35) p<0.001. Conclusion: Newborns of unvaccinated mothers don’t have Pertussis IgG antibodies. This study supports the need of maternal immunization against Bordetella Pertussis.

017
EFFECT OF NONINVASIVE VENTILATORY SUPPORT IN CARDIORESPIRATORY INDICATORS OF PRETERM NEWBORN SUBMITTED TO DIFFERENT BODY POSITIONS

Gabriela Taveira Estrela; Thais Haddad Silveria; Carolina Sano Sugu; Flávia Caetano Rodrigues Tavares Naldi; Marisa Afonso Andrade Brunhoerotti. UNIFRAN

Goal: Evaluate the effect of continuous positive airway pressure in preterm newborn cardiorespiratory indicators in two positions. Methodology: Study was observational, analytical, prospective, randomized and crossover. Thirty-two preterm children, hemodynamically stable in the first 24 hr of life, participated. There were two groups: sixteen premature infants without ventilatory support (Weight 1.789 ± 255g, Gestational age 32.9 ± 1.5) and
sixteen premature infants in noninvasive ventilation by nasal pronga (Weight 1,353 ± 281g, GA 20.9 ± 2.0). The children were allocated in prone and supine positions and cardiorespiratory indicators were observed: respiratory rate, heart rate, oxygen saturation and Silverman Andersen Score (SAS). The indicators were observed by 60 min and recorded every 10 minutes, totaling seven records. The study was approved by the Ethics Committee of the University of Franca. Results: Despite the cardiorespiratory indicators present with expected values for age, children without ventilatory support in supine position responded with higher values in cardiorespiratory and indicators lower oxygen saturation value. The group that was in the noninvasive ventilatory support presented the best results and the values obtained from the cardiorespiratory indicators were close between the positions. On comparison between the groups statistically significant difference was found in the cardiorespiratory variables in the body positions (RR supine and prone, HR prone, oxygen saturation supine and prone, SAS supine) with p = 0.000, among the groups the only variables that showed no statistical difference were the heart rate in the supine position (p = 0.62) and SAS in the prone (p = 0.45). Conclusion: Continuous positive airway pressure collaborates to better cardiorespiratory values, allowing smaller influence of body position. Thus, in children without positive pressure support the supine position showed smaller benefit

Objective: in the present study we aimed to determine whether interleukin-17 (IL-17) gene variation rs2275913 is associated with the severity of acute bronchiolitis (AB). Material and Methods: we recruited infants admitted in the pediatric emergency in the Hospital São Lucas (HSL) PUCRS diagnosed with AB. Infants without hospitalization were recruited at a primary health care center in Porto Alegre/RS. Capillary blood samples were stored using the HapMap database (version 28.0) in order to select the most frequent IL-17 gene polymorphism. The quantification of DNA samples had been performed with DNAhs Qubit Kit (Invitrogen, Carlsbad, CA). Single nucleotide polymorphism (SNP) rs2275913 of the IL-17 gene was genotyped using RT-PCR (TaqMan System, Applied Biosystems, California). The IL-17 polymorphism was screened using the HapMap database (version 28.0) in order to select the most frequent IL-17 genetic variations described in Caucasian populations. Results: 121 cases and 71 controls participated in the final analysis of this genetic association study. Of these, 85.9% and 94.2% of patients had the genotypes GG/AG in the control group and acute bronchiolitis group, respectively. The rare allele of SNP rs2275913 of IL-17 showed a protective effect for severe AB, with higher frequency of homozygous AA (14.1% vs 5.8%; p = 0.047) in the control group (without hospitalization). Moreover, we observed a significant difference when comparing length of hospitalization among patients in the AB group. Conclusion: the study suggests that the polymorphism rs2275913 of IL-17 gene is associated with protection of AB and can influence the severity of AB in these children. This variation may be a marker to identify high-risk patients for AB.

Objective: To evaluate adherence to treatment after educational intervention in patients with Cystic Fibrosis (CF). Material: prospective cohort study. Population: children and adolescents with a diagnosis of CF; the age of six years and followed up at the clinic. Among exclusion criteria motor inability to perform the tests proposed, dependence on oxygen therapy, being in lung transplant list or not complete all meetings. Methods: Patients underwent an educational plan a year in five meetings. The meetings took place parallel to the routine consultation which received information to the patient and held discussion on topics of treatment. At baseline was collected anthropometric data, spirometry and applied: Walk Test of 6 Minutes, instrument of self-reported adherence to treatment, knowledge questionnaire on the management of the disease. Furthermore, the first educational approach was taken: General Aspects of FC. On dates 2, 3 and four themes: Chest physical therapy; Nutrition; Physical activity. At the meeting five was made revaluation. Statistical Analysis, data were expressed as mean and standard deviation and absolute and percentage frequency. Paired t test was performed for pre and post intervention. The level of significance was set at p < 0.05. The statistical software: SPSS 18.0. Results: A sample of 19 children and adolescents with a mean age 10.7 years (± 2.25) and FEV1: 79.57 % (± 25.5). Until now, we have one loss and 15 completed the protocol. After educational plan adherence improved in food (87.5% (±10.9) – 93.6% (±7.4) p < 0.04), physical activity (61.2% (±26.6) – 91.3% (±16.1) p < 0.002) and overall treatment (79.4% (±15.1) – 92.7% (±12.3) p < 0.000). On the management of the disease was not significant. Conclusions: Educational intervention improves adherence to CF treatment in diet and physical activity, as well as the overall treatment of the disease. Limit small sample size.

Aim: Assess the impact of repeated forced spirometric maneuvers on parameters of impulse oscillometric system (IOS) in healthy children and the relation between spirometric parameters and basal oscillometric with those obtained after three maneuvers. Methods: observational and transverse analytic research, including healthy children between six and 12 years, whose healthiness has been proven through questionnaires and normal spirometry (FEV1 and FVC>80% of predicted). Forced spirometric maneuvers were conducted pursuant the American Thoracic Society and considered the parameters FVC, FEV1 and PEF. The maneuvers were performed three times with an interval of 90 sec between them, and to evaluate the impact of these was used IOS (Pneumatográfo Master Scope IOS/Jaeger®, Germany) following each maneuver. The oscillometric parameters were analyzed: ZS, Z5, R20, X5 and FRES. For data analysis, initially there was normality by Kolmogorov-Smirnov test, then applied ANOVA for repeated measures and Friedman test; to relate the Pearson or Spearman test was used. The level of significance was 5%(p < 0.05) in all tests. Results: participated 147 healthy children, 78 girls, with mean age of 9.22(±1.91); and normal spirometry values (mean FVC:95.76%; FEV1:91.24%). When analyzed, there was significant increase of ZS, R5, FEV1 and X5 (p < 0.02) compared with baseline values after each spirometry maneuver. All baseline oscillometric absolute parameter correlated with the absolute spirometric values was significant after three maneuvers. In the
basal spirometric parameters, only the percentage of the predicted FEV1 value had a significant relation with the oscillometric absolute value of X5 (p < 0.001) obtained after the third maneuver. Conclusion: Repeated forced spirometric maneuvers change the IOS parameters verified by progressively increasing the resistance (R5, R20). There is a relationship between baseline oscillometric parameters and spirometric parameters obtained after performing three maneuvers.

021 QUALITY OF SLEEP AND QUALITY OF LIFE IN CHILDREN AND ADOLESCENTS WITH SICKLE CELL DISEASE

Laísa Danielle Alves de Souza Santos (Universidade Federal da Bahia); Tatiane da Anunciação Ferreira (Universidade Federal da Bahia); Silvian Henrique de Souza Miranda (Universidade Federal da Bahia); Renata Maria Pereira Vieira Barbosa (Universidade Federal da Bahia); Vinicius Ramos Machado (Escola Bahiana de Medicina e Saúde Pública); Regina Terse Trindade Ramos (Universidade Federal da Bahia).

Purpose: Verify the relationship between quality of life (QoL) and quality of sleep (QoS) in children and adolescents with sickle cell disease (SCD). Material/Methods: Patients from 06 to 18 years old with SCD without comorbidities and whose parents agreed to participate in the study were invited. Applied the questionnaires: sociodemographic; Pediatric Quality of Life Inventory (PedsQLTM4.0), which assesses QoL; Sleep Disturbance Scale for Children (SDSC) and Epworth Sleepiness Scale (ESS), to evaluate the QoL. Methods: Cross sectional study with descriptive basis and comparison group. Data were collected through structured interviews, with questionnaires including clinical and subjective QoL information, besides PedsQLTM4.0, which comprehends physical, mental, social, educational, psychosocial and global QoL scores, as well as SDSC and ESS, which evaluate the presence of sleep disorders and daytime sleepiness, respectively. For data analysis, descriptive statistics and Pearson correlation analysis (p < 0.05) were used. Results: Evaluated 58 children with SCD and 60 controls. Comparing the SCD and comparison group, the mean age in years was 12.5(±3.1) vs. 11.6(±3.1) (p = 0.10); most were males in the SCD group (p = 0.01); non-white children were the majority in both groups (p = 0.13). Scores obtained from the scales show that: 24.1% of children with SCD and 28.3% of controls had sleep disorder and Respiratory Sleep Disorders (RSD) was the only disorder demonstrated in the cases. Negative and statistically significant correlations were observed between the global score of sleep disorders and the physical, emotional, psychosocial and global subscales from parental reports (rs = -0.5; p < 0.001 for all subscales). Conclusions: It has been shown that children with SCD have worse global QoL when compared to the comparison group and best QoS. Sleep disorders and quality of life subscales showed negative and significant correlations when cases were evaluated.

023 SCREENING FOR CFTR GENE VARIANTS IN PATIENTS WITH CYSTIC FIBROSIS IN NORTHERN BRAZIL.

Valéria de Carvalho Martins (Hospital Universitário João de Barros Barreto – Universidade Federal do Pará); Andrea Kelly Cristina Ribeiro Santos (Universidade Federal do Pará); Ida Vinassa Doederlein Schwartz (Universidade Federal do Rio Grande do Sul).

Aims: To screen for genetic variants associated with cystic fibrosis in a patient population with substantial ethnic heterogeneity. Methods: This is a cross-sectional, outpatient study. All patients were recruited from the CF clinic Hospital University HUJB, and all provided written informed consent for participation. Clinical data were obtained from a retrospective chart review. Peripheral blood samples were collected into EDTA-containing tubes for genomic DNA extraction with the phenol-chloroform technique and quantitation in a NanoDrop 1000 spectrophotometer (Thermo Scientific, Wilmington, DE, USA). Genomic DNA was extracted from 125 cystic fibrosis patients from Northern Brazil. Exons 11, 12, 18, 19, 21, and 22 of the cystic fibrosis transmembrane conductance regulator (CFTR) gene and their exon-intron junctions were sequenced. Results: The sample comprised 125 patients (70 male, 55 female; mean age, 15.4 ± 11.8 years). Ten patients had both pathogenic variants and 22 had only one variant identified. Exon 11 accounted for 77.6% of findings, and a high frequency of the non-pathogenic variant p.M470V (76%) was detected. The p.F508del mutation was found in 21.6% of subjects. The p.G542X mutation was detected in only one patient and the non-pathogenic variant c.3469-65C>G was found in two subjects. We also describe a novel pathogenic mutation (p.T501I). Conclusion: The mutation profile observed in CF patients from Northern Brazil seems distinct from that seen in European populations and in Southern Brazil. The present study highlights the importance of genetic analysis to improve understanding of phenotypic expression and possibly standardize the molecular diagnosis of CF. Clarifying the role of the non-pathogenic variant p.M470V may improve our understanding of the pathophysiology of CF and its phenotypic differences, facilitating identification of genetic origins and ancestry in these groups.

Pediatric Pulmonology
SEVERE ACUTE VIRAL BRONCHIOLITIS IN INFANTS: CLINICAL FEATURES, ETIOLOGY AND OUTCOMES
Alfonso Eduardo Alvarez; Fernando Augusto de Lima Marson; Juliana Santiago; Carmen Silvia Bertuzzo; Clarice Weis Arns; José Dirceu Ribeiro. Universidade Estadual de Campinas (UNICAMP)

Introduction: The minority of patients with Acute Viral Bronchiolitis (AVB) requires hospitalization for oxygen therapy; few studies characterize these infants separately. Objectives: To study and compare clinical features and etiology with the outcomes of infants with severe Acute Viral Bronchiolitis (SABV). Material: Patients hospitalized for SABV in two winter seasons in 3 tertiary pediatric hospitals. Methods: Cross-sectional, prospective cohort study that included all infants with SABV, defined by the need for hospitalization for oxygen therapy, seen in 3 tertiary pediatric hospital in two consecutive winter seasons. Patients were evaluated for epidemiological variables and severity of disease and underwent nasopharyngeal aspirate for detection of virus by PCR technique. Results: 181 infants (176 <1 year) admitted to hospital were included: 58% male, median age 3.8 months (range – 14 to 622 days); median birth weight 3102 grams (range – 565–4850 grams). The frequencies of respiratory viruses identified by PCR in nasopharyngeal aspirates were: Respiratory Syncytial Virus (RSV) 69.5%, RSV-A 52.6%, RSV-B 17.9%, Rhinovirus 26.6%, Coronavirus, Para influenza, Adenovirus, Influenza, Bocavirus, Metapneumovirus and enterovirus had a frequency <5%. Co-infection occurred in 21.4%. Evaluated outcomes were length of stay, median 6.5 days, duration of oxygen therapy, median 5 days, necessity (34%) and duration of ICU stay, median 9 days, necessity (21%) and duration of mechanical ventilation, median 8.5 days. Death occurred in 5 patients (2.8%). The worst outcome correlated positively with: atopy in mother, father and siblings; low maternal schooling, greater number of siblings, larger number of people living in the house and RSV-B (p < 0.05).

Conclusions: The presence of family history of atopy, low maternal schooling, large families and RSV-B were the variables associated with increased severity in the outcomes of SABV.

EFFECT OF THREE METHODS OF Spacer DISINFECTION IN A PEDIATRIC EMERGENCY SETTING
Jussara Aparecida Resende; Eduardo Lima Leite Praça; Amanda Cristina Silva Tardelli Pizzarro; Maria August Aumaral Campos; Wilson Rocha Filho; Chalene Guimarães Soares Mezencio. Hospital Infantil João Paulo II / FHEMIG

Objective: To compare the effectiveness of three different disinfection methods for spacers used in emergency department (ER) of a pediatric hospital. Materials and Methods: This is an open, prospective, longitudinal, randomized study. Twenty four spacers (plastic and metal) used in children admitted to the ER of our institution were randomized to one of three methods of disinfection: 70% ethyl alcohol, 2% sodium hypochlorite or 0.2% per acetic acid. A total of 12 cultures per spacer were obtained. Samples were collected at the end of treatment and after disinfection through swab and washout of mask, chamber and valve. Chocolate agar plate, thioglycolate broth and vitek cards were used for identification and antibiotic susceptibility of microorganisms. Results analysis was made using the chi-square test or Fisher exact test. Results: The spacers were equally distributed in the three disinfection methods. Cultures obtain by swab before disinfection found 14 positive results in the mask (58.3%) and 2 positive results in the chamber (8.3%). There was no bacterial growth from the valve swab. Cultures obtain by washout prior to disinfection found 9 positive results in the mask (37.5%). There was no bacterial growth from the washout of the container or valve. Twenty six microorganisms were identified in 14 of the 24 spacers analyzed. There was no bacterial growth in any spacer after disinfection regardless the method used. Conclusion: Ethyl alcohol at 70%, 2% sodium hypochlorite and 0.2% per acetic acid were equally effective in the disinfection of plastic and metal spacers used in the pediatric ER.

NUTRITION STATUS OUTCOMES OF CF PATIENTS AT DIAGNOSIS AND 12 MONTHS LATER
Janine Prainelli Martins (Universidade Federal de Ciências da Saúde de Porto Alegre – UFCSPA); Gabrielle Carra Forte (Programa de Pós Graduação em Pneumologia – UFRGS); Miriam Isabel de Souza dos Santos Simon (Programa de Pós Graduação em Saúde da Criança e do Adolescente UFRGS Hospital de Clínicas de Porto Alegre); Matias Epifanio (Hospital São Lucas – PUCRS); Leonardo Araujo Pinto (Hospital São Lucas – PUCRS); Paulo Marostica (Programa de Pós Graduação em Saúde da Criança e do Adolescente UFRGS Hospital de Clínicas de Porto Alegre)

Objective: To assess nutritional status outcomes in cystic fibrosis (CF) pediatric patients at diagnosis and 12 months later. Methods: CF patients
followed at the pediatric CF outpatient clinics from Hospital de Clínicas de Porto Alegre and Hospital São Lucas da Pontifícia Universidade Católica do Rio Grande do Sul, in Southern Brazil were evaluated at diagnosis, six and 12 months after, from 2009 to 2014. Generalized estimating equation was used to evaluate nutritional outcomes along time. The association between BMI percentile evolution with clinical and demographic variables was assessed by multivariable linear regression models. Nutritional data were analyzed as percentiles according to the WHO equations. Results: 47 patients were followed in the study. Median age was 0.27 (0.15–1.33) years, and 51.1% were male. Neonatal screening was performed in 30 (63.8%) patients, and pancreatic insufficiency was present in 45 (95.7%) patients, and 19 (40.4%) were homozygous for Phe508del. There were significant increases in mean BMI, Weight/Height, Height/Age and Weight/Age percentiles after 12 months of follow up (55.11 ± 4.18, 31.85 ± 4.14, 53.57 ± 4.46 and 43.4 ± 4.43) compared to the same data at baseline (21.64 ± 3.77, 18.02 ± 3.79, 26.68 ± 4.21 and 15.49 ± 3.45). There was also a significant albumin level increase after one year follow-up (4.34 ± 0.04) compared to baseline levels (3.50 ± 0.15). Neonatal screened patients had a significant higher increase of 31.2% in delta BMI percentile as compared with symptom diagnosed patients. A one percent higher BMI at the diagnosis was associated with 0.6 point lower delta BMI percentile after 12 months. Conclusion: In the present study, there was significant improvement of anthropometric outcomes 12 months after CF diagnosis. The initial BMI percentile and nutritional screening influenced nutritional outcomes.

028
REPERCUSSIONS OF THE SIX MINUTE WALK TEST IN THE PARAMETERS OF IMPULSE OSCILLOMETRY SYSTEM (IOS) IN HEALTHY CHILDREN AND ADOLESCENTS
Maíra Seabra de Assumpção (Universidade Estadual de Campinas (UNICAMP)) e Universidade do Estado de Santa Catarina (UDESC); José Díceu Ribeiro (Universidade Estadual de Campinas (UNICAMP)); Renata Maba Gonçalves (Universidade do Estado de Santa Catarina – UDESC); Antonio Manoel Goulart Neto (Universidade do Estado de Santa Catarina – UDESC); Janaina Cristina Scalco (Universidade do Estado de Santa Catarina (UDESC)); Camila Isabel Santos Schivinski (Universidade do Estado de Santa Catarina – UDESC)

Objective: To verify the repercussions of the submaximal exercise test in the respiratory mechanics of healthy children and adolescents. Material: All participants were submitted to evaluation of respiratory mechanics using a Jaeger MasterScreen Impulse Oscillometry System (IOS) (Erich Jaeger, Germany) following American Thoracic Society (ATS) standards. The participants took the six-minute walk test (6MWT), which was carried 2 times with an interval of 30 minutes, respecting the standards of ATS. Methods: Observational cross-sectional analytical study with healthy students aged 7 to 14. Approval by the Ethics Committee of the Santa Catarina State University and consent form were met. Biometric data were evaluated (weight, height, body mass index). The evaluation IOS and spirometry was performed before the first 6MWT (PRE), and repeated immediately after the first (POST1) and second 6MWT (POST2). The data normality verification was realized for Shapiro-Wilk test. The ANOVA (Post-hoc Bonferroni) for repeated measures and Friedman test was applied for comparison. The significance level was set at 5% (p < 0.05). Results: 19 subjects participated, 52.6% boys, and average age of 10.89 ± 2.3. We found differences between the parameters of total airway resistance (R5) and central airway resistance (R20) (p = 0.041 and p = 0.025, respectively). By post-hoc-analysis, we found a significant increase of R5 between the PRE and POST1 (R5: 0.54 ± 0.11 kPraLs/Ls vs 0.59 ± 0.15 kPraLs/Ls, p = 0.013 and R20: 0.44 ± 0.80 kPraLs/Ls vs 0.47 ± 1.0 kPraLs/Ls, p = 0.028). The only spirometric variable that showed significant change was the forced expiratory flow at 25–75% (FEF 25–75%) (p = 0.003), with a significant drop between PRE, POST1 and POST2 (85.95 ± 19.97% vs 80.87 ± 20.25% p = 0.013 and 85.95 ± 19.97% vs 77.91 ± 19.40%, p = 0.004). Conclusions: The repercussions found were the increase in the total and central airway resistance and a reduction in FEF 25–75% after the 6MWT in healthy children and adolescents.

029
THE IMPACT OF PEDIATRIC CHRONIC KIDNEY DISEASE ON LUNG FUNCTION, FUNCTIONAL CAPACITY AND QUALITY OF LIFE
Paulo André Freire Magalhães; Carolina Guimarães Teixeira; Bárbara Bernardo Rinaldo da Silva; Lívia Barboza Andrade; Maria do Carmo Menezes Bezerra Duarte.
Programa de Pós-graduação em Saúde Materno Infantil, Instituto de Medicina Integral Prof. Fernando Figueira

Objective: despite the scientific and technologic improvements in the pediatric renal management, a large proportion of children and adolescents with Chronic Kidney Disease (CKD) still to progress with high morbidity and mortality. Researches regard prevention and early diagnosis of CKD need to be encouraged and may contribute to better outcomes. The aim of the present study was to evaluate the impact of the CKD on quality of life, respiratory muscle strength, lung function, and functional capacity in children and adolescents. Methodology: cross-sectional study of children with CKD aged 8 to 17 years. Pediatric Quality of Life Inventory (PedsQLTM), muscular strength, pulmonary function tests and the 6-minute walking test (6MWT) were applied. Student’s t-test, ANOVA, and Pearson’s coefficient of correlation were used for statistics. Significance was set at 5%. Results: Of the 40 patients, the mean distance walked at the 6MWT was 396 meters, and the mean final score at the PedsQLTM by the children and parents was 50.9 and 51, respectively. From the children’s perspective, transplanted patients had a higher quality of life score when compared to those undergoing hemodiagnosis (p < 0.001); those who practiced physical activity had better score when compared to the sedentary children (p < 0.001). From the children’s and the parents’ perspectives, the male gender had a higher quality of life score (p < 0.05). There was a positive correlation between the distance walked at the 6MWT and age, height, final PedsQLTM, forced vital capacity (FVC), and forced expiratory volume in the first second (FEV1), as well as a negative correlation between FEV1/FVC and the distance walked. Conclusion: significant reduction in the quality of life and the functional capacity was observed in children with CKD, influenced by the type of treatment, gender, and sedentary lifestyle.

030
EVALUATION THE THERAPEUTIC RESPONSE IN THE USE OF THREE FORMS OF MUCUS REGULATORS (HS AND RHDNASE)
Eduardo Augusto Caldeira Storti (Hospital Pequeno Príncipe); Paulo Cesar Kassek (Hospital Pequeno Príncipe); Carlos Antônio Riedi (Hospital Pequeno Príncipe); Silvia Eniko Shimakura (Hospital de Clínicas).

This study aimed to evaluate the therapeutic response in the use of three forms of mucus regulators (Hypertonic Saline-HS and rhDNase), by pulmonar function test and Cystic Fibrosis Clinical Score Kanga (CFCS Kanga).

Methods: 18 CF children aged 6–19 years (male10/female8), was included. All patients used rhDNase and HS in three schemes: Daily rhDNase, rhDNase on alternate days associated with HS daily, and rhDNase on alternate days. Each treatment lasted 8 weeks, totaling 24 weeks. There was no interval between treatments. All patients received the medication by jet nebulizer (PARI Proneb®): rhDNase (Pulmozyme® 2.5 mg / 2.5 ml) and HS 7% twice a day plus salbutamol MDI. At the start and at the end of the 8-week treatment period, each patient was submitted to an interview and a physical exam to fill the CFCS Kanga. The ECFs Kanga measures the correlation of signs/symptoms (cough, appetite, sputum, dyspnea, energy) with objective medical assessment (temperature, respiratory rate, auscultation lobes and weight). In each treatment, patients and caregivers were asked...
to give a score of their symptoms using a score from 1 to 3 (1 improvement, 2 indifferent, 3 worsening). Results: None of the medications caused severe adverse effects. The respiratory microbiology wasn’t altered between the schemes. The median baseline FEV1 and FEF 25–75 were, respectively, 88.45% and 55.05% of predicted, and during the treatments, no significant statistical difference was observed between the forms of treatment. By CFCS score, the treatments differed from baseline values (p<0.05) but they do not differ between them (P>0.8). Patients and caregivers preferred rhDNase on alternate days associated with HS on a daily scheme as the best in symptoms improvement. Conclusion: All forms of treatment showed improvement in respiratory symptoms, however, for the CF treatment, rhDNase every other day associated with HS appeared to be more efficient.

031 NIGHT CONTINUOUS SATUROMETRY IN NEWBORNS WITH SUSPECTED SLEEP APNEA

Natalia Galaz Souza (Universidad de Concepción); Ximena Andrea Paulette Navarro Tapia (Hospital Guillermo Grant Benavente); Daniel Zenteno Arazas (Hospital Guillermo Grant Benavente); Aldo Bancalari Molina (Hospital Guillermo Grant Benavente); Pablo Brockmann Veloso (Red Salud Pontificia Universidad Católica de Chile).

Objectives: Sleep studies in newborns (NB) are usually been used to guide therapeutic conducts; in this regard, Polgraph (PG) is a diagnostic method that has reference values recently reported. Night continuous saturometry (CSO?) is a test widely used in neonatology; however, its diagnostic yield is not known for these patients. We aim to determine the diagnostic performance of the CSO? in RN with suspected sleep apnea. Materials and patients: 39 patients were included. Between January 2013 and June 2015 simultaneous results of CSO? and PG from infants hospitalized with patients: 39 patients were included. Between January 2013 and June 2015 performance of the CSO? in RN with suspected sleep apnea. Materials and methods: All patients with a clinical diagnosis of CF attending our outpatient pediatric clinic were invited to enroll in the study. Informed consent was given by the parents and informed assent by patients older than 6 years old. Total DNA was extracted from blood samples, quantified and submitted to purification. Ion Ampliseq HiFi mix kit (Life technologies) was used for library preparation and next generation sequencing was performed on the IonTorrent PGM platform (Life Technologies). Results: 126 CF patients were enrolled and 42 relevant mutations were identified in 244 alleles, 6 of which had not been described yet. Mutations c.580-2A>C (712-2A>C), c.1084_1088dup (S364insTATGTA), c.2476G>T (E826X), c.3728T>A (L1243X) and c.4168delC (4300delC) were all identified in patients with a classic CF-phenotype who also had another deleterious mutation present. The c.785C>T (Y262I) mutation was identified in a CF patient along with two other CF-causing mutations. Conclusions: Mutations 712-2A>C, S364insTATGTA, 4300delC, E826X and L1243X are very likely pathogenic since these are splice-site, frameshift and nonsense mutation and were identified in CF patients with another CF-causing mutations. The missense mutation Y262I is most likely a neutral mutation, not affecting CFTR function, since it was identified in a patient with 2 other CF-causing mutations.

033 NUTRITIONAL STATUS, NUTRITIONAL MONITORING AND FOOD SECURITY IN CHILDREN BELOW 18 YEARS, DIAGNOSED WITH CYSTIC FIBROSIS IN ANTIOQUIA COLOMBIA

Cristian Sepúlveda (Universidad de Antioquia); Catalina López (Universidad de Antioquia); Olga Morales Márquez (Universidad de Antioquia y Hospital San Vicente Fundación); Tiffanni S. Tobón (Universidad de Antioquia); Adriana M. Zea (Universidad de Antioquia); Iván D. Florez (Universidad de Antioquia).

Aim: to determine the nutritional status and feeding practices of children with Cystic Fibrosis (CF) registered in the Mariana Pro-Fibrosis Quística Foundation in Antioquia, Colombia. Methodology: This was a cross-sectional study. Anthropometric measures were taken, the nutritional classification was based on the Cystic Fibrosis Foundation (CFF) parameters of the United States and it was compared with the World Health Organization (WHO) standards. In addition, the food security in the children’s households was assessed. Variables associated with malnutrition were explored using chi2 test. Results: we evaluated 67 patients with CF, 56.7% were female, mean age 8.94 years (SD 4.63). Based on CFF, the proportion of children with acute and chronic malnutrition were 44.4% and 47.8%, respectively. According to WHO they were 15% and 38.8%, respectively. The proportion of households with food insecurity was 34.3%. The 38.8% of the patients had no regular clinical monitoring by nutritionist. The lack of monitoring by nutritionist was associated statistically significant with acute (P = 0.019) and chronic malnutrition (P = 0.027) based on the CFF criteria. A higher number of hospitalizations was statistically significant in patients with acute malnutrition (P = 0.016) and chronic malnutrition (P = 0.027) using WHO criteria.

Conclusions: Acute and chronic malnutrition were higher that reported in the literature for pediatric patients with CF. Following the WHO criteria we classified less children with malnutrition than with the CFF criteria. The lack of monitoring by nutritionist was the most associated factor with acute and chronic malnutrition according to the CFF criteria. Acute and chronic malnutrition is a factor which is related with increased need of hospitalizations. A proper nutritional intervention could improve the nutritional status in children with Cystic Fibrosis.

034 ABSENCE OF SEASONAL EFFECT IN THE CONTROL OF ASTHMA IN CHILDREN FROM SOUTHERN BRAZIL

Pediatric Pulmonology
Objective: To evaluate the seasonal effect on the control of asthma in children from an outpatient tertiary clinic from Southern Brazil. Methods: We have included children with asthma (mild, moderate and severe disease), aged between 7–17 years, who were followed for 12 months. Spirometry, control of disease, adherence to treatment, body mass index (BMI) and physical activity, during the four seasons of the year (spring, summer, autumn and winter), between 2012 and 2013, were assessed. Results: We have included 35 patients (mean age: 10.63 ± 2.088), with 17 (48.6%) male subjects. Twelve children were classified as mild (34.3%), 12 as moderate (34.3%) and 11 as severe (31.45%) asthma. There were no differences between seasons of the year and the outcomes evaluated, except for the use of controller medications, with 100% (n=35) of patients receiving controller treatment in the winter and 76.5% (n=26) in the summer (p=0.028). Nineteen (65.5%) subjects reported to be nonadherent to treatment. Spirometry tests (FEV1, FVC, FEV1/FVC) were similar between seasons (p=0.582, 0.616 and 0.846, respectively). Similar results for control of disease, physical activity levels, and BMI were observed between seasons. In addition, when severity of disease was stratified (mild, moderate and severe asthma), no outcome differences between seasons were detected. Conclusion: Children with asthma used less controller treatment during summer time, but did not show any improvement of disease during this period. Seasonal asthma behavior should be better understood and strategies of clinical preventive treatment during summer time should be reviewed some populations.

035 QUALITY OF LIFE AND LUNG FUNCTION IN CF PATIENTS USING TOBRAMYCIN INHALATION POWDER (TIP)

Maristela Trevisan Cunha (Instituto da Criança HCFMUSP); Regina CTP Juliani (Instituto da Criança HCFMUSP); Claudio Leone (Departamento Materno Infantil Saúde Pública); Joaquin C Rodrigues (Instituto da Criança HCFMUSP); Fabiola V Adde (Instituto da Criança HCFMUSP)

Objectives: to evaluate the domains of the Cystic Fibrosis Questionnaire-Revised (CFQ-R), FEV1, the peak inspiratory flow (PIF) and the inhalation time pre and post 3 cycles of TIP. Patients’ opinion about TIP was also inquired. Patients and methods: open-label, prospective study, included CF patients 6 to 20 years with FEV1 above 25%, chronic Pseudomonas aeruginosa infection, previous treatment with tobramycin inhalation solution (TIS). Written informed consent was obtained. CFQ-R, spirometry, PIF and inhalation time were evaluated at day 1 and day 29 of the 1st and 3rd cycle of TIP (T0, T1, T2 and T3, respectively). At T3 the patients were asked what they found about TIP and what was their preference (TIS or TIP). Statistical analysis: a sample size of 24 patients was estimated to have 80% power to detect a difference of at least 8 points in the treatment burden domain of the CFQ-R with an alpha of 5%. Friedman test was used to compare the values at T0, T1, T2 and T3. Results: 24 patients, 10 female, 13.9 ± 3.4 years were evaluated with median FVC of 74%, FEV1 of 55.5% at T0 with no significant changes throughout the study. Significant changes occurred in the median PIF at T0xT1 (85 ± 95L/min; p = 0.02) and in the TIP inhalation time at T0xT3 (10 ± 4 minutes, p < 0.0001). Median CFQ-R domain scores at T0 were: respiratory 77.8; treatment burden 77.8; physical 81.2; emotional 81.6; social 71.8, body image 94.4; eating 88.9; digestive 100; role 100; vitality 83.3; health 88.9; weight 83.3. Significant changes occurred in the median PIF and inhalation time were evaluated at day 1 and day 29 of the 1st and 3rd cycle of TIP. There was a significant increase in PIF and a decrease in inhalation time. Although the improvement in quality of life measured by the CFQ-R occurred in only two domains all patients preferred TIP.

036 ASTHMA MANAGEMENT RESPONSE: A GENETIC POINT OF VIEW

Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertazzo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Aduleya Dalho Controro Toro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Maria Helena Gonzalez de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Tania Kawasaki de Araujo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Jose Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas).

Objective: To verify the association of allergic asthma (AA) control (uncontrolled, partly controlled or controlled) with polymorphisms (pol) in genes related with the inflammatory response. Material: Included 205 AA patients, aged between 6–18 years, and 536 healthy controls aged over 18 years with no history of personal and/or family of lung illness in 2 generations. Methodology: Asthma control was measured based on international guidelines. 256 polymorphisms (pol) on 125 genes were considered for association study. For genetic analysis, OpenArrayTM platform (LifeTechnologies®) was used. SPSS® v22.0 performed the statistical analysis. Alpha = 0.05. Results: Of the 205 AA patients, asthma was uncontrolled in 49 (23.9%), partly controlled in 22 (10.7%) and controlled in 132 (65.4%). 68 genes (110 pol) were associated with the degree of AA control, comparing each group with healthy control subjects. Uncontrolled AA: 38 genes were associated (5 were uniquely associated — GIPC3, IL12B, PSMC6, RORC and TBX21). Odds ratio (OR) range: 11.86 (95%CI = 3.61–38.96) for rs35094768*C in TBX2AR gene to 0.054 (95% CI = 0.007–0.39) for rs4986791*CT in TLR4 gene. Partially controlled AA: 23 genes were associated (3 were uniquely associated — CTNNB1, IL12R1 and NFG). OR range: 16.59 (95%CI = 2.61–9.63) for rs6136*GG in SELP gene to 0.005 (95%CI = 0.007–0.39) for rs4986791*CT in TLR4 gene. For controlled asthma: 54 genes were associated (22 were uniquely associated – B9D2, CHR1, IL10, IL18R1, IL1RL1, IL33, IL4, ILRAK, ITGB5, JAK1, LRRC, LAT4H, NFKBIB, NOD1, PDE4D, SOD2, STAT6, TFAP2A-2AS1, TIMD4, TGFBI, TSLP and ZBTB). OR range: 5.559 (95%CI = 3.17–9.74) at rs35094768*C in TBX2AR gene to 0.095 for rs4880*GG (OR = 0.01–0.70) in SOD2 gene. Conclusion: AA control was associated with variants studied. In the future, the AA management may be directed by knowledge of basic genetic profile that acts in AA variability and could provide better quality of life for the patients.
adherence was 50% (CI 95% 39.7–60.3, p < 0.01). We found a lower risk of lack of medication adherence in patients with severe and moderate asthma compared with patients with mild asthma; OR 0.27 (CI 95% 0.09 to 0.77) and OR 0.63 (CI 95% 0.44 – 0.91) respectively. We also found a lower risk of lack of medication adherence in patients with poorly controlled compared to patients with controlled asthma and in patients whose caregivers considered appropriate to use inhaled steroids. Conclusion: The study found a high rate of lack of medication adherence in asthma similar to reported in other studies, and a statistically significant association between severity, disease control and the perception of the use of inhaled steroids, with this phenomenon.

038 PERFORMANCE OF THE PAEDIATRIC ASTHMA QUALITY OF LIFE QUESTIONNAIRE IN BRONCHIOLITIS OBLITERANS

Angela de Moura (Pontificia Universidade Catolica do Rio Grande do Sul); Edgar E. Savria (Universidade de Santa Cruz); Suellen Goecks Oliveira (Pontificia Universidade Catolica do Rio Grande do Sul); Helena T. Mocelin (Universidade Federal de Ciencias da Saude de Porto Alegre); Gilberto Bueno Fischer (Universidade Federal de Ciencias da Saude de Porto Alegre); Rita Mattiello (Pontificia Universidade Catolica do Rio Grande do Sul).

Aims: To assess the performance of Paediatric Asthma Quality of Life Questionnaire (PAQLQ) in patients with Post-infectious Bronchiolitis Obliterans. Method: Children and adolescents aged 8 to 17 years old and with previous diagnosis of Post-infectious Bronchiolitis Obliterans (PBO) were enrolled. They are all followed up periodically at the pediatric pulmonology outpatient clinics of Hospital da Criança Santo Antonio, in Porto Alegre, Brazil. Spirometry followed recommendations from the ATS/ERS. The PAQLQ has 23 items items distributed in three domains: Activity Limitation, Symptoms and Emotions with similarly answered by means of a 7-point likert scale, from 1 (severely affected) to 7 (unaffected). The Psychometric Properties assessed were reliability and validity. Reliability was measured using the Cronbach's alpha coefficient (CAC) and ceiling and floor effect for individual domain and overall PAQLQ scores. Construct validity was evaluated using Pearson correlation coefficients between PAQLQ scores and VEF1 (percent of predicted). The study was approved by the institutional Review Board. Results: A total of 41 children, 33 (81%) boys, mean age 11.9 ±2 (years) was evaluated. Mean values of FEV1 was 48.3 ± 17.6. The mean PAQLQ total score was 5.05 ± 1.5, the lowest score was for the domain “activity limitations”, and highest (5.18) for the “emotion” domain; The PAQLQ exceeds the reliability (CAC) criterion of 0.90 recommended for analyzing individual patient scale scores for all domains and total score (0.92 IC95% 0.869 to 0.956). There were no significant floor or ceiling effects in questionnaire. Correlations between FEV1 and instrument were r = 0.062, P = 0.763. Conclusion: The PAQLQ showed good performance in children with Post-infectious Bronchiolitis Obliterans. The results demonstrate that children with PBO can reliably and validity self-report their HRQOL with an asthma specific disease instrument.

039 A NEW INSIGHT ON CFTR ALLELE FREQUENCY IN BRAZIL THROUGH NEXT GENERATION SEQUENCING

Luiza Mesquita Nunes (Centro de Pesquisa Experimental, Hospital Israelita Albert Einstein); Roberto Ribeiro (Instituto de Medicina Tropical, Universidade de São Paulo); Ester Sabino (Instituto de Medicina Tropical, Universidade de São Paulo); Vivian D. T. Niewiadomski (Diagnostico das Américas S. A); Guilherme Lopes Yamamoto (Instituto da Criança, Universidade de São Paulo); Luiz Vicente Ribeiro F. da Silva Filho (Instituto da Criança, Universidade de São Paulo); Centro de Pesquisa Experimental, Hospital Israelita).

Background: As of 2013 only 40% of patients in the Brazilian CF Registry had any kind of genetic investigation, and more than half had at least one allele with no mutation identified. The use of mutation panels tailored to European and North American populations might not be adequate to investigate Brazilian population due to its ethnic admixture. Better knowledge of allele frequency in our population is essential towards developing more efficient screening tools for CFTR mutations. Objectives: To sequence all the exons in the CFTR gene of patients attending our CF center in São Paulo, Brazil, and compare the allele frequency found with Brazilian Registry data. Methods: All CF patients attending our pediatric clinic were invited to enroll. Informed consent was given by the parents and assent by patients older than 6 years old. Total DNA was extracted from blood samples, quantified and submitted to purification. Library preparation was performed using Ion Ampliseq HiFi mix kit (Life Technologies) and sequencing using the Ion Torrent PGM platform (Life Technologies). Results: 126 CF patients were enrolled and 32 deleterious mutations were identified in 244 alleles. The 3 most common mutations were: c.1521_1523delCTT (F508del), c.1624G>T (G542X) and c.2988 + 1G>A (3120 +1G>A) with, respectively, 145 (60,2%), 18 (7,3%) and 13 (5,3%) alleles. While the two first were also the most frequent in the Brazilian Registry, there were marked differences in the frequency of other mutations, such as 3120 +1G>A, mostly found in patients of African descent, with only 13 (0,5%) alleles in the Registry. Conclusions: There is a striking difference in the frequency of mutations found among our patients when compared to the Registry data, probably due to the diagnostic approach adopted in the routine so far. In the era of the growing concept of personalized medicine, there is a need to expand genetic investigation of CF patients to other Brazilian regions.

040 CYSTIC FIBROSIS: CLINICAL OUTCOMES OF PATIENTS WITH GASTROSTOMY

Raquel Maccarenhas Freitas; Marcela Duarte de Sillos; Sonia Mayumi Chiba; Beatriz Neuhaus Barbisan; Juliana Ferreira Mauri da Silva; Cleviss Eduardo Tadeu Gomes. Universidade Federal de Sao Paulo

Objective: To describe the clinical outcomes of patients with cystic fibrosis (CF) after percutaneous endoscopic gastrostomy (PEG). Methods: We reported anthropometric and clinical data from patients followed in the CF multidisciplinary clinic who underwent PEG. Data were obtained just before PEG placement and at 6, 12 and 24 months after the procedure. Results: Six patients underwent PEG, four female and two pancreatic sufficient. Median age was 13.5 years (13 to 17 years). They were all colonized by Pseudomonas aeruginosa. Clinical indication of PEG was malnutrition without response to hypercaloric oral diet. Caloric intake varied from 1700 to 2660 kcal per day. Before the PEG, five patients were wasted and one was severely wasted. Three had low height for age. After 6 and 12 months of the procedure, five adolescents had a better z-score in the BMI. One of them died due to pulmonary complications. The 6th patient did not gain weight in the first year after the procedure due to non-adherence. After 24 months of the procedure, three patients (n = 5) became eutrophic. The non adherent patient improved his adherence in the 2nd year and, despite of not reaching eutrophy, increased his BMI. Another adolescent, who had obtained a good result in the 1st year, because of worsening of respiratory symptoms in the 2nd year, showed intolerance to diet infusion and had a reduction in the BMI z- score. Regarding to height, after 24 months, all of them improved and reached height for age z-score. It was observed an important increase in the caloric intake of all patients, ranging from 2800 to 5500 kcal per day. There were no serious complications in the immediate or late postoperative period. Conclusion: In most patients, nutritional therapy via PEG was efficient and did not induce severe complications. The PEG acceptance by the patient and his family was essential to the procedure success. The evolution of the respiratory condition cannot be separated from the nutritional one.
041
RELATION BETWEEN RESPIRATORY MUSCLE STRENGTH AND PHYSICAL ACTIVITY IN HEALTHY SCHOOL.
Rafaela Coelho Mantsky; Fabiula Joana da Mata Belem; Francieli Camila Mucha; Patrícia Morgana Rentz; Keil; Camila Isabel Santos Schivinski. Universidade do Estado de Santa Catarina

Aim: Relate respiratory muscle strength and level of physical activity in healthy children as well as comparing the force between active and sedentary children. Materials and Methods: observational and transverse analytic research, quantitative character. Healthy children were included from private and public schools of Florianópolis (SC / Brazil). The health has been proven through questionnaires, as well as spirometric parameters, respecting the guidelines of the American Thoracic Society. The level, frequency and intensity of regular physical activity was assessed through physical activity questionnaire for children PAQ-C, representing the very sedentary interval (1) to very active (5). In accordance with this instrument, the groups were divided in sedentary (with score 1 and 2) and active (with score 3, 4 and 5). The evaluation of respiratory muscle strength was conducted through the manometer MVD 300 (Globalmed49, Brazil), for which were obtained data inspiratory maximum pressures (MIP) and expiratory (MEP). Data analysis began with the Kolmogorov-Smirnov normality test and then applied the Spearman correlation test. For comparison between active and sedentary groups, the Mann-Whitney test. The level of significance was 5% (p <0.05) in all tests. Result: They attended by 131 children, 69 girls, with a mean age of 9.28 (± 1.87). The group of sedentary children, was composed of 32 children, including 22 girls; active children were 99 children, 53 boys. The mean of MIP was 79.30 (± 29.42) x 86.03 (± 38.24), and MEP of 80.62 (± 23.48) x 86.08 (± 20.44), respectively. When evaluated parameters showed no correlation. When comparing the averages between the two groups, there was no significant difference. Conclusion: Respiratory muscle strength is not related to the level of physical activity in healthy children as well as comparison between active and sedentary children was not significant.

042
TREATMENT ADHERENCE IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS IN ANTIOQUIA (COLOMBIA)
Olga Lucía Morales Márquez (Universidad de Antioquia y Hospital San Vicente Fundación); Diana Arocha Hoyos (Universidad de Antioquia); Ana Gómez Gómez (Universidad de Antioquia); Javier Contreras Ortiz (Universidad de Antioquia).

Introduction: Treatment adherence is a major issue for patients with cystic fibrosis (CF), since they require complex and chronic treatment that needs dedication of the patients and relatives. Objective: To measure the prevalence of lack of treatment adherence and explore its associated factors in children and adolescents with CF diagnosis. Methods: Descriptive cross-sectional study. A structured survey about treatment routine and the main barriers to adherence was carried out in 48 patients or relatives. Lack of treatment adherence and disease severity were measured. Results: Prevalence of lack of adherence in the sample using the Morinsky Green test was 62.5%; related barriers were: little interest in treatment, lack of time, forgetting to take medication and problems with health insurance. Conclusion: The study identified a high prevalence of lack of treatment adherence in a sample of children and adolescents from Antioquia – Colombia with CF-diagnosis, but analysis of related barriers showed that any of them were statistically significant, although this analysis was limited by the small sample size.

043
COMPARATIVE ANALYSIS OF THE CONCENTRATIONS OF CHLORIDE AND SODIUM IN SWEAT AND SALIVA OF PATIENTS WITH CYSTIC FIBROSIS AND CONTROLS
Aline Cristina Gonçalves (Graduate Program in Child and Adolescent Health, Department of Pediatrics, School of Medical Sciences); Antonio Fernando Ribeiro (Department of Pediatrics, School of Medical Sciences of Unicamp and Pediatric Gastroenterology Labor); Eliete Ap Lomau (Department of Pediatrics, School of Medical Sciences of Unicamp and Pediatric Gastroenterology Labor); José Direceu Ribeiro (Department of Pediatrics, School of Medical Sciences of Unicamp and Pulmonary Physiology Laborator); Andre Moreno Morcillo (Department of Medical Clinic School of Medical Sciences of Unicamp); Carlos Emilio Levy (Department of Clinical Pathology, School of Medical Sciences of Unicamp and Division Clinical Pathol).

Objectives: To evaluate the use of saliva as a diagnostic fluid for Cystic Fibrosis (CF). Materials and Methods: Saliva samples were collected simultaneously and sweat of 163 patients [CF (n= 67) and no CF (n= 96)]. The saliva samples were collected with a roll-shaped cotton – Salivette® (Sarsted-Germany – http://www.sarstedt.com) for three minutes, after fasting for 1 hr and embrocation with gauze moistened with deionized water or mouthwash with drinking water. The samples were immediately centrifuged at 1500 rpm for 15 minutes, amounts less than 0.6 mL were discarded. Chloride concentration were analyzed by ABL gasometer (model 835, Radiometer®, Denmark) by gasometric analysis using an ion-selective electrode direct. was performed according to Gibson and Cooke. Results: The mean age For the CF group (CFG) was 10.25 years and for control group(CG) was 4.20 years old. Sweat chloride: For CFG the of the was SD 32.33 mmol/L, mean = 118.65mEq/L and median = 121mEq/L, already in the CG the SD was 7.95 mmol/L, mean = 17.53 mEq/L, median = 14.39 mEq/L. Saliva chloride: In CFG SD = 18.17 mmol/L, mean = 31.82 mmol/ L, median = 26 mmol/L and in CG: SD = 9.94 mmol/L, mean = 18.06 mmol/L and median = 17 mmol/L. Sweat sodium: CFG in the SD = 31.59 mEq/L, mean = 98.50mEq/L, median = 95.67mEq/L and in CG: SD = 9.26 mEq/L, mean = 16.58 mEq/L, median = 13.91mEq/L. Saliva sodium: CFG the SD =11.44 mmol/L, mean = 21.8 mmol/L, median = 18 mmol/L and in CG the SD = 7.45, mean = 15.16, median = 14 mmol/L. Comparing concentrations of chloride and sodium in the of CFG sweat with the group of patients non CF p <0.001, as well as in saliva. The Cut-off saliva for chloride was 19.5 mmol/L. Conclusion: As well as in sweat, in saliva also the concentrations of chloride and sodium ions are significantly elevated in CF patients, when compared to individuals non CF, which enables that there is the possibility use of the dosage of chlorine and sodium ions in the saliva for the diagnosis of CF.

044
PERFORMANCE OF INFECTION/COLONIZATION CHRONIC OF THE LUNG IN CYSTIC FIBROSIS: A GENETIC MODEL TO ESTABLISH THE CLINICAL VARIABILITY
Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertacza (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Adyleia Dalbo Contrera Toro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Antonio Fernando Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Direceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas).

Objectives: To investigate Pseudomonas aeruginosa (PA) [mucoid(MPA), non-mucoid(NMPA) and first isolation of PA(1PA)] – bigger severity: >729 months, and minor severity: 29 months]. Achromobacter xylosoxidans (AX), Burkholderia cepacia(BC) and Staphylococcus aureus(SA) prevalence in cystic fibrosis(CF) patients with two CFTR mutations identified and genes associated with inflammatory response. Material: Included 126CF patients with 2CFTR mutations identified (classes I, II and/or III). Pediatric Pulmonology
046 ASSOCIATION BETWEEN NUTRITIONAL STATUS, BY NUTRITIONAL RISK SCREENING TOOL, LUNG FUNCTION IMPAIRMENT AND MORBIDITY IN CYSTIC FIBROSIS PATIENTS (OR CHILDREN AND ADOLESCENTS)

Andressa Brancher Roeder (Universidade Federal de Santa Catarina); Daniela Barbieri Hauschild (Programa de Pós Graduação em Nutrição); Yara Maria Franco Moreno (Programa de Pós Graduação em Nutrição); Luiza Kasulke de Lima (Universidade Federal de Santa Catarina); Renata Cardoso Escobar Ferreira (Universidade Federal de Santa Catarina); Eliana Barbosa (Departamento de Nutrição e Dietética – Hospital Joana de Gusmão).

Aims: Evaluate the association between the nutritional status of children and adolescents with cystic fibroses (CF) during a nutritional risk screening tool (NRST), lung function and the clinical outcomes after 1 year of follow-up. Material: Children and adolescents with CF, aged from 6 to 15 years old, clinically stable, at a CF treatment centre in southern Brazil. Methodology: Prospective cohort study, developed from July 2013 to December 2015. At the baseline (t0), demographic, clinical data, NRST and lung function were recorded. The NRST, developed specifically for CF children, included data regarding weight gain, growth rate and current body mass index. Lung function was evaluated by forced expiratory volume in 1 sec (FEV1). After 1 year of follow up, lung function, Pseudomonas aeruginosa (PA) infection and hospitalization were collected from medical records. Logistic regression was applied and p<0.05 considered as significant. Results: At baseline (t0) 46 patients were assessed, with a median age of 8.5 years, 47.8% were female, 51.3% with VEFI < 80% and 30% DF508 Homozygote. Twelve patients (26.1%) were classified as low risk, 17 as moderated risk (36.8%) and 17 as high risk (36.8%). After 1 year, 48.4 % had VEFI <80%, 27.3% presented PA infection and 23.6% were hospitalized. The patients classified as moderated risk presented an odds ratio (OR) 6.12 for lung function impairment <80% (Confidence Interval (CI) 95% 0.83; 45.02), OR 0.95 (CI95% 0.13; 7.22) for PA infection and, OR 0.64 for hospitalization (CI95% 0.08; 5.42). The patients classified as high risk showed an OR 4.2 of lung function impairment (CI95% 0.59; 30.09), OR 1.94 PA infection (CI95% 0.29; 13.19) and (OR) 2.7 for hospitalization (CI95% 0.43; 16.94). Conclusion: Although no association between NRST, lung function and the clinical outcomes was observed, the results suggest that the NRST can be a useful tool to evaluate the nutritional status of patients with CF.

047 FUNCTIONAL AND IMAGE FINDINGS OF POST-INFECTION BRONCHIOLITIS OBLITERANS FROM LATIN AMERICA COHORT (BOLAT)

Edgar E. Sarria Icaza (Universidade de Santa Cruz do Sul); Rita Mattiello (Pontificia Universidade Católica do Rio Grande do Sul); Helena T. Mocelin (Universidade Federal de Ciências da Saúde de Porto Alegre); Gilberto Bueno Fischer (Universidade Federal de Ciências da Saúde de Porto Alegre).

Aim: To describe functional and image findings of Post-infectious Bronchiolitis Obliterans (PBO) from Latin America cohort. Methods: The study included patients with diagnosis of PBO and undergoing treatment in pediatric pulmonology outpatient clinics in six countries: Brazil, Chile, Argentina, Peru, Paraguay and Uruguay. Patients were diagnosed following clinical and radiological criteria. Centers followed

Pediatric Pulmonology
International recommendations for acceptability and reproducibility criteria for spirometry maneuvers. Computed tomography scans and spirometry were retrieved from hospital records for this study. Results: We retrieved data from 696 children with PIBO, 551 male (73%), mean age 10 years. The median times for diagnosing was 7 months, and for hospitalization at diagnosis was 30 days. The median of pre-BD values for all parameters were clearly lower than that of reference values, especially FEV25-75% (FVC = 63%) (IQ47-79); FEV1 = 51% (IQ 41-61); FEV1/FVC = 39% (IQ18-60); FEF25-75% = 18% (IQ1-27). Main computed tomography findings from 435 images were Hyperinflation 348 (78%); Mosaic pattern 282 (65%); Bronchiectasis 301 (67%); Atelectasis 279 (63%). Conclusions: The analysis of clinical and functional characteristics of Post-infectious bronchiolitis obliterans from Latin America cohort (BOLAT) confirms a pattern of severe pulmonary function and structural impairment, characterized by marked airway obstruction and presence of diffuse lung damage.

048 FUNCTIONAL CAPACITY ASSESSMENT (TGLITTRE – P) IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS: BEHAVIORAL IMPULSE OSCILLOMETRY PARAMETERS

Ana Carolina da Silva Almeida (Hospital Infantil Joana de Gusmão / Universidade do Estado de Santa Catarina); Francieli Camila Mucha (Universidade do Estado de Santa Catarina); Renata Gonçalves Maba (Universidade do Estado de Santa Catarina); Maira Seabra de Assumpção (Universidade do Estado de Santa Catarina); Gustavo David Ludwig (Universidade Federal de Santa Catarina); Camila Isabel dos Santos Schivinski (Universidade do Estado de Santa Catarina).

Introduction: assessment of functional capacity in patients with cystic fibrosis (CF) monitors the progress of the disease and the effect of therapeutic interventions. Thus, the impact of TGlittre-P has not been studied in CF, specifically in parameters of impulse oscillometric system (IOS). Objective: Analyze the behavior of IOS parameters of children/adolescents with CF after performing the TGlittre-P. Materials and Methods: Cross-sectional observational analytical survey conducted in Florianópolis/SC – Brazil. Attended by children/adolescents clinically stable. Led to the examination of IOS (Pneumotachograph Jaeger Master Scope IOS/Germany) according to the norms of the American Thoracic Society, before and immediately after the completion of the TGlittre-P, being considered for analysis the parameters: Z, R5, R20, X5, Fres and AX. TGlittre-P was conducted two times, at an interval of 30 minutes, being considered the best performance test. The test consists of loading a backpack during the course of a circuit that involves multiple tasks. Assign a better performance in the circuit conducting five laps in the shortest time possible. After checking the data normality the Shapiro-Wilk test, we applied the Wilcoxon test, with significance level of 5% (p < 0.05) (software SPSS 20.0 software). Results: Participated 21 children/adolescents, 12 girls between 6 and 15 years of age (mean: 9.59 ± 2.27) and mean BMI of 16.51 ± 3.35 kg/m². They had a mean percentage of predicted FVC: 85.52 ± 19.80% and FEV1: 68.47 ± 21.70%. Increased by all the oscillometric parameters after performing the TGlittre-P, but without statistical significance (p > 0.05), as described: Z: 8.74 ± 2.72 × 9.18 ± 2.94 cmH2O/L/s; R5: 5.18 ± 2.43 × 8.62 ± 2.74 cmH2O/L/s; R20: 5.11 ± 1.17 × 5.84 ± 1.2 cmH2O/L/s; X5: 3.03 ± 1.38 × 3.05 ± 1.31 cmH2O/L/s; Fres: 23.06 ± 4.9 × 23.65 ± 5.35 (1/L); AX: 29.22 ± 20.76 × 29.02 ± 22.99 cmH2O/L. Conclusion: The realization of TGlittre-P for children/adolescents with stable CF did not change oscillometric parameters in this population.

049 COMPARISON OF PIMAX VERSUS SNIFF TEST MANEUVERS TO EVALUATE RESPIRATORY MUSCLE STRENGTH IN CHILDREN WITH NEUROMUSCULAR DISEASE

Natalia Gaião Souza (Universidade de Concepção); Daniel Zenteno Araos (Hospital Guillermo Grant Benavente); Ximena Andrea Navarro Tapia (Hospital Guillermo Grant Benavente).

Objectives: A sniff is a short voluntary inspiratory maneuver performed through one or both unoccluded nostrils. It involves a contraction of the diaphragm and other inspiratory muscles. It is easier to perform than mouth pressure, and it does not require mouth muscles strength. This study aims to evaluate respiratory muscle strength in children with different neuromuscular diseases comparing mouth pressure and nostril pressure. Materials and Patients: 24 patients were included. 13 boys and 11 girls, ages 5–19 years old. Diagnosis: Duchenne Muscular Dystrophy (10), Mietemonegenece (4), Poliartromatizada (1), Escleriosis (5), Sindrome de Goldenhar (1), Atrofia espinal tipo II (1), Ehler Dalos (1), Miopatia (1). We used the MicroRPM digital respiratory pressure meter, rubber flanged mouthpiece, nasal probes in four sizes (extra small, small, medium, large) and a nose clip. Methodology: 24 patients were evaluated, 2 patients were excluded for being unable to perform de mouth pressure maneuver. Patients performed a maximal inspiratory pressure maneuver, rested 30 minutes, and afterwards performed a sniff test. Results: All of the patients were able to perform sniff test, 2 were unable to perform mouth pressure maneuver. Sniff pressure was significantly higher than Pimax (p < 0.05). Conclusions: Sniff test was feasible to perform in all patients regardless age and diagnosis. The value for respiratory muscle strength was underestimated when evaluated with Pimax compared to Sniff test in children with neuromuscular disease.

050 TGLITTRE-P IN CHILDREN AND ADOLESCENTS WITH CF: BEHAVIOR OF PHYSIOLOGICAL VARIABLES

Ana Carolina da Silva Almeida (Hospital Infantil Joana de Gusmão / Universidade do Estado de Santa Catarina); Francieli Camila Mucha (Universidade do Estado de Santa Catarina); Renata Gonçalves Maba (Universidade do Estado de Santa Catarina); Suelen Bittencourt Rosa (Universidade do Estado de Santa Catarina); Gustavo David Ludwig (Universidade Federal de Santa Catarina); Camila Isabel dos Santos Schivinski (Universidade do Estado de Santa Catarina).

Introduction: AVD test Glittre proposed to evaluate functional capacity in COPD, has been adapted for pediatric (TGlittre-P), but there is no evidence of its impact in cystic fibrosis (CF). Aim: To analyze the behavior of physiological variables in TGlittre-P performed by children / adolescents with CF. Materials and methods: cross-sectional observational analytical study involving patients with CF monitored in Florianópolis / SC-Brazil, whose clinical stability was guaranteed by the application of two clinical scores. Patients were characterized by anthropometry and spirometry (Pneumotachograph Jaeger Master Scope IOS/Germany), conducted under the American Thoracic Society. Following, there was the TGlittre-P, which consists of loading a backpack along the way by a task multi circuit (up and down stairs, sitting and standing, walking, and move objects). Performance relates to go five times in this circuit in the shortest time possible. TGlittre-P was performed 2 times at 30 min intervals. The physiological variables: (heart frequency (hf), and breathing frequency (bf), blood pressure systolic (bps) and diastolic, pulse oxygen saturation, and dyspnea) were observed before and immediately after the test. Statistical data normality was conducted by test Shapiro-Wilk and applied to the Wicolxon with significance level of 5% (p < 0.05) (software SPSS® 20.0). Results: participated 21 children / adolescents between 6 and 15 years (mean: 9.59 ± 2.27 years), 12 girls. Mean BMI was 16.51 ± 3.35 kg/m² and spirometric parameter predicted FVC was 85.52 ± 19.80% and FEV1 68.47 ± 21.70%. After testing, a significant increase (p < 0.05) the hf (94.62 ± 28.03 ± 131.1 ± 23.54 bpm), bf (25.71 ± 4.72 ± 32.14 ± 6.606 ppm) and bps (90.71 ± 14.51 ± 96.9 ± 18.33 mmHg), no significant change in other physiological variables. Conclusions: TGlittre-P by children / adolescents with CF has increased significantly hf, bf, and bps.
S22 Abstract

051 VOLUMETRIC CAPNOGRAPHY AS A VENTILATORY EVALUATION INSTRUMENT IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS, SUBMITTED TO A SIX MINUTES SUBMAXIMAL STRESS TEST

Paloma Lopes Francisco Parazzi (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Maria Angela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Camila Isabel Santos Schivinski (State University of Santa Catarina, Center of Physical Education and Sports); Antônio Fernando Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas).

Objective: Volumetric Capnography (VCap) is studied as a form of VE, partial pressure of CO2 in arterial blood (VCO2), dead space to tidal volume ratio (VD/VT) and end-tidal carbon dioxide pressure (PetCO2). The analysis takes place before, during and after the submaximal stress test. Before the test, the variables showed a difference in value as well, although only statistically significant for these indicators: spirometry (p < 0.001), SpO2 (p < 0.001), Respiratory frequency (p < 0.001), VCO2 (p < 0.001), VE/VCO2 (p < 0.001), PetCO2 (p < 0.001), Borg scale (before the test: p > 0.001, and after the test: p > 0.005). Conclusion: The VCap is an instrument that may be used for ventilatory parameters analysis during physical exercise. All cardiopulmonary variables from spirometry and capnography showed difference in patients with CF when compared to healthy individuals before and after physical exercise.

052 CHILDREN WITH HIGH RISK OF SEVERE RESPIRATORY SYNCTIAL VIRUS INFECTION: PREVALENCE AND RISK FACTORS FOR RECURRENT WHEEZING

Maria Cristina Ribeiro Dos Santos Simões: Natasha Yumi Matsunaga; Maria Angela Gonçalves De Oliveira Ribeiro; José Dirceu Ribeiro; André Moreno Morcillo; Adylene Aparecida Dalbo Contrera Toro. UNICAMP

Objective: To assess the prevalence and risk factors associated with development of recurrent wheezing in children with high risk for severe Respiratory Syncytial Virus infection and to analyze if the passive immunization modified the prevalence of recurrent wheezing. Methods: This cross-sectional study was conducted between June 2014 and August 2015. A questionnaire was administered to the parents of the children studied. Group I received passive immunization with Palivizumab whereas Group II did not receive the drug. Results: Five hundred and twelve infants were analyzed with mean age 39.5 (±5.64) months. In Group I, 261 children with a mean gestational age 31.2 (±4.94) weeks and median 29 (24–41) weeks. Group II, 251 preterm children with a mean gestational age 34.0 (±1.79) and median 34 (28–36) weeks, p < 0.001. The prevalence of recurrent wheezing among the 512 children was 25.8% (95% CI: 22.1–29.7). In Group I, the prevalence was 30.7% (95% CI: 25.3–36.5) and in Group II, 20.7% (95% CI: 16.0–26.1). These results showed a statistical significant difference (p = 0.011). Risk factors associated with recurrent wheezing by univariate analysis were: birth weight less than 1500g; gestational age below 32 weeks; lack of breastfeeding; hospitalization for wheezing; living with more than three children in the same household; and atopy in children and parents. With multivariate analysis, the associated independent factors were: gestational age below 28 weeks (aOR: 6.52; 95% CI: 2.55–16.69; p < 0.001); food allergy in children (aOR: 2.92; 95% CI: 1.45–5.86; p = 0.003); atopic dermatitis in children (aOR: 2.61; 95% CI: 1.37–4.97; p = 0.004) and parental asthma (aOR: 1.76; 95% CI: 1.09–2.85; p = 0.020). Conclusion: The prevalence of recurrent wheezing was 25.8%. There was statistical difference in the prevalence of recurrent wheezing between the two groups. Gestational age below 28 weeks, food allergy, atopic dermatitis and parental asthma were defined as associated risk factors.

053 SEVERE ACUTE VIRAL BRONCHIOLITIS AND ALLERGIC ASTHMA: A POSSIBLE GENETIC RESPONSE COMMON

Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertuzzo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Maria Angela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Andressa de Oliveira Peixoto (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Alfonso Eduardo Alvarez (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas).

Objectives: To determine the common genetic profile for the risk and severity for allergic asthma (AA) and severe acute viral bronchiolitis (SAVB) by genes related to inflammatory response. Material: Included 186 SAVB patients with clinical diagnosis aged between 0–12 months, 221 asthma allergic (AA) patients and 536 healthy controls aged over than 18 years with no personal and/or family history of lung illness in 2 generations. Methodology: 256 polymorphisms (pol) on 125 genes were considered for association study. For genetic analysis, OpenArray™ platform was used. SPSSv22.0 performed the statistical analysis considering the percentage of bronchodilator response. ? = 0.05. Results: AA was associated with 56 genes (86 pol). SAVB was associated 51 genes (65 pol). AA + SAVB was associated with 60 (86 pol). As risk factor, the odds ratio amplitude (ORA) was 8.82 (95% CI = 5.833 to 13.33; rs35094768° AC, TXB2AR gene) and 7.52 (95% CI = 4.436 to 12.74; rs6334° AG, NTRK1 gene) to 1.35 (95% CI = 1.014 to 1.799; rs597980° AG, ADAM33 gene); (95% CI = 1.005 to 1.808; rs4697177° GG, KCNIP4 gene) and in Group II, 20.7% (95% CI:16.0–26.1). These results showed a statistical significant difference (p < 0.020). Conclusion: The prevalence of recurrent wheezing was 25.8%. There was statistical difference in the prevalence of recurrent wheezing between the two groups. Gestational age below 28 weeks, food allergy, atopic dermatitis and parental asthma were defined as associated risk factors.

Pulmonology
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ASSOCIATION BETWEEN HYDRATION STATUS AND CLINICAL OUTCOMES IN CYSTIC FIBROSIS PATIENTS AFTER ONE YEAR FOLLOW-UP

Laiza Kosulke de Lima (Universidade Federal de Santa Catarina); Andresa Brancher Roeder (Universidade Federal de Santa Catarina); Renata Cardoso Exobaru Ferrreira (Universidade Federal de Santa Catarina); Daniela Barbieri Hauschild (Programa de Pós Graduação em Nutrição); Yara Maria Franco Moreno (Programa de Pós Graduação em Nutrição); Elaina Barbosa – Departamento de Nutrição e Dietética – Hospital Infantil Joana de Gusmão).

Aims: to evaluate the association between hydration status, evaluated by vectorial analysis of bioelectrical impedance (BIVA), lung function and morbidity in children and adolescents with cystic fibrosis (CF). Material: Children and adolescents with CF; aged 6 to 15 years old, clinically stable, conducted in a CF treatment center from South Brazil. Methodology: Proropositive cohort study. At baseline there were collected weight, height, bioelectrical impedance (BIA) parameters and lung function. For BIVA, resistance and reactance vectors were collected and, the patients were classified as hyperhydrated (HH), normally hydrated (NH) and dehydrated (DH). After 1 year, lung function, hospitalization and Pseudomonas aeruginosa (P. aeruginosa) infection were registered from medical records. Forced expiratory volume in 1 sec as percentage of predicted (FEV1) was assessed to measure lung function. A logistic regression was applied with p < 0.05 as significant. Results: There were recruited 46 CF patients, median age of 8.5 years, 52.5% male and 30% DFS580 homozygote. Among these 45.6% were NH, 13.1% HH and 41.3% DH. After 1 year, 48.4% had FEV1 < 80%, 27.8% were infected by P. aeruginosa and 23.8% were hospitalized at least once. The patients classified as DH presented an odds ratio (OR) of 4 [Confidence Interval (CI) 95% 0.80; 20.02] for FEV1 < 80%, 1.67 (CI95% 0.33; 8.37) for P. aeruginosa infection and 3.10 (CI95% 0.64; 15.00) for hospitalisation. HH patients presented an OR of 2 (CI95% 0.21; 18.69) for FEV1 < 80%, 2.50 (CI95% 0.30; 20.45) for P. aeruginosa infection and 1.13 (CI95% 0.09; 13.44) for hospitalization. Conclusion: There were not observed an association between BIVA and the outcomes. The use of BIVA as a nutritional tool is promising once it considers the hydration status. More studies with a larger sample needed to confirm the applicability of BIVA in CF patients.

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ANTHROPOMETRIC AND BIOCHEMICAL MARKERS AS PREDICTORS OF LUNG FUNCTION IN CYSTIC FIBROSIS PATIENTS

Gabrielle Carra Forte (Universidade Federal do Rio Grande do Sul); Miriam Isabel de Souza dos Santos Simon (Hospital de Clinicas de Porto Alegre, Programa de Pós Graduação em Saúde da Criança e do Adolescente); Paulo José Cauduro Marostica (Hospital de Clinicas de Porto Alegre, Programa de Pós Graduação em Saúde da Criança e do Adolescente)

Objective: to evaluate anthropometric and biochemical markers as predictors of lung function in cystic fibrosis (CF) patients. Methods: cross-sectional study with 78 CF patients aged 6 to 19 years, followed at the Hospital de Clinicas de Porto Alegre CF outpatient clinics. To collect data, a structured questionnaire with gender, age at diagnosis, pancreatic insufficiency, bacterial colonization, albumin, spirometry and anthropometric data were filled. All data were collected from electronic medical records. A multivariate linear regression model was built to evaluate the relationship between biochemical, clinical and nutritional variables and spirometry. Results: Seventy nine patients were enrolled in the study, 39 (50%) were females. The mean age was 12.8 ± 3.8 years and the median age at diagnosis was 1.32 (0.4–6.0) years. Sixty (76.9%) patients were colonized by Staphylococcus aureus (Sa), 42 (53.8%) by Pseudomonas aeruginosa (Pa), 15 (19.2%) by mucoid Pa, 9 (11.5%) by methicillin resistant Sa (MRSA), and 19 (24.4%) by Burkholderia cepacia complex (Bc). The mean albumin was 4.2 ± 0.4 mg/dL and the mean predicted forced expiratory volume in one second (FEV1%) was 81.9 ± 22.6%. Mean body mass index (BMI) percentile was 49.9 ± 27. Patients with lower than 25 BMI percentile had a 12.5% lower FEV1%. An albumin increase of 0.1 mg was associated with a 2.72% increase in FEV1% predicted, and one year increase in age was associated with a reduction in 1.22% of FEV1% predicted. Conclusion: BMI percentile, albumin and age were independent predictors of FEV1% in a tertiary referral hospital. The results show the importance of assessing biochemical and anthropometric markers, together with lung function in CF patients.

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IS RESPIRATORY MUSCLE STRENGTH CRITICAL FOR OBTAINING THE SPIROMETRIC MANEUVER SLOW VITAL CAPACITY IN HEALTHY CHILDREN?

Rafaela Coelho Minsky; Gabriel Silvestrin; Francieli Camila Mucha; Patricia Morgana Rentz; Keil; Maylli Daiani Graciosa; Camila Isabel Santos Schivinski Universidade do Estado de Santa Catarina

Aim: To analyze if there is a relationship between respiratory muscle strength (RMS) and the ability to do slow spirometry maneuvers, as well as compare the strength of healthy children who can and those who can’t perform these maneuvers. Materials and Methods: Observational and transverse analytic research with the participation of healthy children.
between 6 and 12 years. Health questionnaires were administered to confirm the healthiness as well as spirometry (Pneumatógrafo Jaeger – Master Scope IOS®, Germany). According to the American Thoracic Society (FEV1 e FVC>80% do predico). Based on this test, it is considered that children who were able to perform the slow spirometry maneuvers – reaching 80% predicts – were the first group, and those who have not reached that percentage formed the group 2. Further to the RMS was measured (MVD manometer 300 Globalmed®, Brazil) to obtain data inspiratory maximum pressures (MIP) in expiratory pressure (MEP). After application of the Kolmogorov-Smirnov normality test, analyzed the Spearman correlation to relate the RMS values with SVC and IC, and the Mann-Whitney test to compare the MIP and MEP between the groups. It is considered a significance level of 5% (p<0.05). Results: participated in 83 children with a mean of 8.54 years (± 1.95). Group 1 was consisting of 44 children, 25 girls and showed a significant relationship between spirometric variables and FMR (p<0.005). The same relation was observed in group 2 comprised 39 children (20 girls) (p<0.005). Group 1 showed higher values of MIP (81.70 ± 26.13 1 ± 49.46 ± 28.05cmH20) and de MEP (86.43 ± 24.61 1 ± 64.44 ± 18.34cmH20) compared to group 2, respectively, Conclusion: There was a significant relation between the RMS values and the ability to do slow spirometric maneuvers, and the RMS of children who can perform the maneuvers is greater than those that can’t.

059 FACTORS ASSOCIATED WITH SEVERE ACUTE VIRAL INFECTIONS OF THE LOWER RESPIRATORY TRACT IN A POPULATION OF HOSPITALIZED CHILDREN

Angela Maria Pedroza (Hospital Clinica San Rafael, Bogota); Carlos Rodriguez (Universidad Nacional De Colombia); Rainnery Acuna (Universidad Militan Nueva Granada)

Summary: Introduction Viral acute lower respiratory infections (ALRIs) are important causes of morbidity and mortality, especially among children under 5 years of age living in low- and middle-income countries (LMIC). Severe ALRI poses a significant health problem because of its great healthcare and clinical burden. Although predictors of severe ALRI in children have been reported, there have been few research studies performed in LMIC. Objectives. The aim of the present study was to determine predictors of disease severity in a population of Colombian children <5 years of age with ALRI. Methods. In a prospective cohort study, we determined independent predictors of severe ALRI in a hospitalized population of children under 5 years old with ALRI during a 1-year period (2014–2015). We included both underlying disease conditions and the infecting respiratory viruses as predictor variables of severe disease. We defined severe disease as the necessity of pediatric intensive care unit admission. Results. Of a total of 1180 patients admitted with a diagnosis of ALRI, 416 (35.3%) were selected for the present study because they were positive for any kind of respiratory virus. After controlling for potential confounders, it was found that a history of pulmonary hypertension (RR 3.62; CI 95% 2.38–5.52; p<0.001) and a history of recurrent wheezing (RR 1.77; CI 95% 1.12–2.79; p=0.015) were independent predictors of severe disease. Conclusions. The present study shows that respiratory viruses are significant causes of ALRI in infants and young children in Colombia, a typical tropical LMIC, especially during the rainy season, which is defined as the 3-month period from March to May in the country. Additionally, the results of the present study show that clinical variables such as a history of pulmonary hypertension and a history of recurrent wheezing are more relevant for predicting ALRI severity than the infecting respiratory viruses.

060 IMPULSE OSCILLOMETER SYSTEM (IOS), SPIROMETRY AND SECONDHAND SMOKE IN HEALTHY CHILDREN AND ADOLESCENTS

Pediatric Pulmonology

Maíra Seabra de Assumpção (Universidade Estadual de Campinas (UNICAMP) e Universidade do Estado de Santa Catarina (UDESC)); Fernanda Cabral Xavier Sarmento de Figueiredo (Universidade do Estado de Santa Catarina – UDESC); Josué Dícoro Ribeiro (Universidade Estadual de Campinas); Renata Mabu Gonsalves (Universidade do Estado de Santa Catarina – UDESC); Letícia Godart Ferreira (Universidade Estadual de Campinas – UNICAMP); Camila Isabel Santos Schivinski (Universidade do Estado de Santa Catarina – UDESC)

Objective: To verify the repercussions of the exposure to secondhand smoke (SS) in the impulse oscillometry and spirometry parameters in healthy children and adolescents. Material: All participants were submitted to evaluation of respiratory mechanics and lung function using a Jaeger MasterScreen Impulse Oscillometry System (IOS) (Erich Jaeger, Germany) following American Thoracic Society (ATS) standards. Methods: Comparative cross-sectional study with healthy students aged 6 to 14. Biometric data were evaluated (weight, height, body mass index) as predictors and for sample characterization. The evaluation IOS (three tests were recorded, with data acquisition for at least 20 sec) and spirometry was performed. The sample was divided according to the exposure report to SS: exposed group (SSG) and no history of exposure (NSSG). For data normality verification, the Kolmogorov-Smirnov test was applied, and T-test or Mann-Whitney to compare spirometric and oscillometric parameters between the groups (p<0.05). Results: 78 children and adolescents participated (39 group 1, 14 boys and 25 girls in each group). There was difference in mean values of parameters peak expiratory flow (PEF) (p=0.01) and forced expiratory flow between 25 and 75% of vital capacity (FEF25-75%) (p=0.04) and forced expiratory volume in one second in percentage FEV1% (p=0.05) in SSG showing lower average values than NSSG. SSG presented has higher absolute average values in IOS, including the parameters of total airway resistance: R5 (p=0.02) and indicators of the presence of airway obstruction: ZS, p=0.03; resonant frequency: Fres, p=0.03 e reactance area: AX, p=0.01). Conclusion: The children and adolescents exposed to SS had lower values of spirometric variables and increased oscillometric variables. Changes in the function and mechanic respiratory were identified in the SSG

061 EFFECT OF OVERWEIGHT AND ASTHMA ON LUNG FUNCTION IN CHILDREN

Marcus Herbert Jones; Morgana Fernandes; Cristian Roncada; Paolo Pizze; Renato Stein PUCRS

Objectives: The objective of this study is to analyze the effect of asthma and body mass on lung function in children. Methodology: Asthmatic children and healthy controls were recruited in public and private schools in Porto Alegre, Brazil. Spirometry, before and after bronchodilator was obtained in the Lung Function Laboratory (PUCRS) at the same time weight and height were measured. Lung function and body mass index were transformed to z-scores for all analysis using international equations. Results: 188 children (97 girls, 52%) were enrolled, 114(60%) with asthma and 85(45%) overweight. The mean and standard deviation of age, height and weight were 11.1 ± 1.1, 146.7 ± 8.4 and 44.2 ± 12.3 respectively. There was a positive correlation between BMI and FVC and FEV1 and a negative correlation with FEV1/FVC. Children with asthma had lower FEV1/FVC and FEF25-75 and higher FVC (p<0.05 for all). Overweight children had higher FVC and PEV1 and lower FEV1/FVC (p<0.05 for all). Stratifying the analysis by asthma (yes/no) and BMI (normal weight/overweight) we found significant differences between overweight asthmatic children and overweight/non-overweight controls for FEV1/FVC and FEF25-75 (AN-OVA, p<0.01 for all comparisons). Overweight asthmatic children also had lower PEV1/FVC when compared to non-overweight asthmatic children (p<0.01 for all). There was a significant trend for higher FVC and lower FEV1/FVC and FEF25-75 in overweight children with asthma. (Jonckheere-Terpstra, p<0.01 for all analysis). Conclusion: Our findings suggests a worsening of lung function in asthmatic children who are overweight
expressed by reduction in FEV1/FVC and FEF25-75. The concomitant increase in FVC and FEV1 suggests that the accelerated somatic growth may have affected the chest or lung development. The findings of this study suggest that being overweight during childhood promotes a dysanaptic pattern of lung growth and that overweight asthmatics may have a high risk for COPD later in life.

062 INTERLEUKIN-8 GENE VARIATIONS AND THE SUSCEPTIBILITY TO SEVERE BRONCHIOLITIS

Lidiane Alves de Azevedo Leitão; Magáíl Mocellin; Ana Paula Duarte de Souza; Leonardo Araújo Pinto.
Centro Infant, Instituto de Pesquisas Biomédicas, FAMED, PUCRS

Objective: almost all infants present infection by respiratory syncytial virus (RSV) up to 2 years of age. However, the severity of acute bronchiolitis (AB) can vary significantly. This variability may be caused by genetic and/or immunological factors. Previous studies have shown that RSV-infected airway contain high levels of interleukin 8 (IL-8). The knowledge of genetic polymorphisms associated with severe AB may have clinical relevance, identifying patients with high-risk for severe bronchiolitis. The aim of the present study was to compare the frequency of IL-8 polymorphisms in infants with severe AB and controls recruited in a low-income area in Southern Brazil. Material and methods: we included infants admitted to the pediatric emergency of Hospital Sao Lucas (HSL) da PUCRS with AB, aged less than 12 months, in the period between 2009 to 2011. Infants of the same age who did not have had AB, were recruited from the primary care center Born Jesus (controls). Capillary blood samples were collected and DNA was extracted for genotyping of single nucleotide polymorphism using FTA Bom Jesus (controls). Capillary blood samples were collected and DNA was extracted for genotyping of single nucleotide polymorphism using FTA Bom Jesus (controls). Genotyping of two polymorphisms (SNPs rs2227543 and rs2227307) in the IL-8 gene was performed using TAQMAN (Applied Biosystems – AB, Carlsbad, California). Results: in the final genetic association study, we included 115 cases and 64 controls. There was no significant deviation from Hardy-Weinberg equilibrium. The SNP rs2227543 showed significant protection for AB, with higher frequency of homozygous TT patients in the control group (OR 0.25: CI 0.10 to 0.65). However, rs2227307 showed no association with AB. Conclusion: this finding, together with the analysis of previous studies, suggests that the IL-8 polymorphism rs2227543 is associated with protection of AB and influences the severity of the disease.

063 ONE-YEAR FOLLOW-UP OF INFANTS HOSPITALIZED FOR SEVERE ACUTE VIRAL BRONCHIOLITIS

Alfonso Eduardo Alvarez; Fernando Augusto de Lima Marson; Juliana Santiago; Carmen Silvia Bertazzo; Clarice Weis Arns; José Dirceu Ribeiro.
Universidade Estadual de Campinas (UNICAMP)

Introduction: Few studies have been conducted to clinically monitor the outcomes after discharge of infants with acute viral bronchiolitis (AVB) that were classified as severe (SA VB). OBJECTIVES: To evaluate and compare clinical and laboratory variables with the outcomes during one year follow-up of patients hospitalized for AVB. MATERIAL: Patients hospitalized for AVB in two winter seasons in 3 tertiary pediatric hospitals. METHODS: All patients with SA VB, defined by the need for hospitalization for oxygen therapy, in two winter seasons in 3 tertiary pediatric hospitals were evaluated for epidemiological variables and severity of disease. Patients underwent nasopharyngeal aspire for detection of virus by PCR technique. All patients were followed-up for one year. Results: 85 patients (< 1 year) were followed-up. Outcomes after 3 and 12 months of discharge (in percentage) were: wheezing (58.8 and 46.1), use of bronchodilators (56.0 and 47.4), use of oral steroids (38.8 and 43.4), need to seek the emergency room (38.1 and 35.1), need of hospitalization (8.3 and 6.4), pneumonia (10.6 and 3.8), 36.5% and 11.8% of patients used inhaled steroids and montelukast, respectively. Characteristics associated with worse outcome were: household smoking, presence of atopic dermatitis, family history of atopy (FHA), low gestational weight, low birth weight, cardiopathy, low maternal schooling, greater number of siblings, larger number of people living in the house, age at admission, length of ICU stay, duration of oxygen therapy and BVA caused by Respiratory Syncytial Virus (RSV) A, RSV B, Rhino virus or by more than 1 virus (p < 0.05). CONCLUSIONS: Infants with SAVB have high rates of symptoms after hospital discharge and need to be followed. Household smoking, atopic dermatitis, FHA, low gestational and birth weight, cardiopathy, low maternal schooling, large families, age at admission, severity of BVA and type of virus were associated with maintenance of symptoms.

064 EVALUATION OF THE VENTILATORY EFFICIENCY INDEX (VE/VCO2) AS A PROGNOSIS MARKER IN PATIENTS WITH CYSTIC FIBROSIS

Paloma Lopes Francisco Parazzì (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Maria Angela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Camila Isabel Santos Schivinski (State University of Santa Catarina, Center of Physical Education and Sports); Antônio Fernando Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas).

Objectives: Physical exercise have been studied as a tool to assess the worsening or the improvement of clinical markers of cardiorespiratory function. Clinical markers of lung function deterioration in cystic fibrosis (CF) have been studied and the relation between ventilation (VE) and the production of carbon dioxide (VE/VCO2) slope can be used to determine ventilatory responses to exercise. Our objective was to evaluate if the VE/VCO2 ratio is a useful marker as a prognostic indicator for children and adolescents with CF. Material: A controlled, clinical trial, with 128 individuals, 64 with CF and 64 in the control group(CG), of both genders, from a University Hospital. Methodology: All individuals completed the submaximal stress test (SST) on the treadmill for 6 min and other exams, after the approval of the Institutional Ethics Committee and signed Informed Consent form. The comparison between the groups of patients with CF and the CG was made through Kruskal-Wallis and Mann-Whitney tests. Results: The analysis of the VE/VCO2 values and VE/VCO2 showed a negative correlation between the two variables, in all time points analyzed; before (p < 0.0001), during (p < 0.0001), and after the SST (p = 0.0017), for both the CF group and the CG. The VE/VCO2 ratio showed significant statistical difference before, during and after the stress test, in both groups (p < 0.0001 for all 3 time points analyzed). The analysis of the VE/VCO2 ratio also showed a difference between genders (p<0,0001) and age groups (from 6 to 12 and 13 to 25; 0,0001). Conclusion: The values of the VE/VCO2 index were higher in the CF group. In patients with chronic pulmonary disease the reasons for these high values are attributed to increased dead space, ventilation and perfusion; therefore showing that the VE/VCO2 is of prognostic value for children and adolescents with CF.

065 STEP TEST IN CYSTIC FIBROSIS

Talitha Di Martha Chacon Belotti; Vanessa Cristina Wiegis Pires de Godoy; Cinthia Johnston; Gustavo Antonio Moreira; Beatriz; Neuhaus Barbisan; Sonia Mayumi Chiba; Universidade Federal de São Paulo UNIFESP

Objective: The aim of this study was to report correlation between the step test (ST) and demographic, clinical and spirometric variables in patients
with CF. Methods: All subjects were attending the pediatric CF clinic at Universidade Federal de São Paulo. They were required to be at least 7 years old in order to be able to execute spirometry and the ST. Spirometry was performed according to Brazilian guidelines, using CPFS/Dusb Medgraphics Spirometer. ST was performed in 6 min and, according to the severity of the disease, reduced to 3 minutes. It was used a 15cm step, according to Balfour-Lyn (1998). We evaluated heart rate (HR), percutaneous oxygen saturation (SpO2), breathlessness, and fatigue of legs before and after each step test. HR and SpO2 were evaluated through a portable pulse Oximeter (mark Moriiya, MD300E model), breathlessness and fatigue of legs through the modified Borg scale. The test performance was measured by the number of steps taken (complete cycle of ascent and descent). Comparison between quantitative measures were performed using Pearson correlation analysis (p < 0.05). Results: We analysed 22 patients, mean age 16 years (∆±4.6), 13 males, 14 were eutrophic, 5 were thin and 2 were severely thin. Mean ± SD of percent-of-predicted FEV₁ was 66.8 ± 29.4%. We found significant correlations between age and FEV₁ (r = -0.55), and between number of steps and FEV₁ (r = -0.45). Number of steps was not correlated with age, BMI or Sw三项man-Kulczycki score. Also, there was not any correlation between Delta of SpO2 and FEV₁. Conclusion: Exercise capacity assessed by the step test showed to be lower as FEV₁ decreases in CF. Other expected correlation resulted negative maybe due to small sample size and the fact that ST is a submaximal test for most subjects and non-progressive.

066 RELATIONSHIP BETWEEN OSCILLOMETRIC AND SPIROMETRIC VARIABLES OF PATIENTS WITH CYSTIC FIBROSIS

Ana Carolina da Silva Almeida (Hospital Infantil Joana de Gusmão / Universidade do Estado de Santa Catarina); Francieli Camila Mucha (Universidade do Estado de Santa Catarina); Renata Gonçalves Maba (Universidade do Estado de Santa Catarina); Mônica Lisboa Chang Wahys (Hospital Infantil Joana de Gusmão); Gustavo David Ludvig (Universidade Federal de Santa Catarina); Camila Isabel dos Santos Schivinski (Universidade do Estado de Santa Catarina).

Introduction: Single pulmonary function measurements are useful to assess the extent of the abnormality, the progression of the disease and the individual response to therapy in cystic fibrosis (CF). Among the evaluation methods of the respiratory system, the most widely used is spirometry. A further embodiment for this type of evaluation is the impulse oscillometric system (IOS), which performs measurements of the mechanical properties of the lung and thorax. Objective: To investigate the relationship between the oscillometric and spirometric variables of CF patients. Methods: Cross-sectional observational analytical survey conducted in Florianópolis/SC- Brazil. Participated clinically stable children and adolescents, according to two clinical scores (CFCS and 11 signals). It was first performed IOS and subsequently spirometry, to ensure that forced spirometry maneuvers did not interfere in the results of oscillometry. Both were carried out with the Pneumotachograph Jaeger Master Scope IOS / Germany, under the rules of the American Thoracic Society, being considered for the analysis of parameters FEV₁, FVC, FEV₁/FVC, Z, R5, R20, X5, Freq and AX. Statistical analysis was conducted using SPSS 20.0 software. There was data normality the Shapiro-Wilk test and applied the Spearman correlation test. The significance level was 5% (p < 0.05). Results: there was a significant relationship between oscillometric and spirometric variables in relation to the percentage of predicted FVC with X5, and percentage of predicted FEV₁ with variables:X5, Freq e AX (p < 0.05). In terms of absolute values there was a significant FVC relationship with: Z, R5, X5, Freq e AX (p < 0.05). Also showed up FEV₁/FVC relationship with X5 e AX (p < 0.05). Conclusion: Oscilometric variable X5 and AX correlated with spirometric parameters relevant in assessing obstructive pulmonary disease (FVC and FEV₁).

Pediatric Pulmonology

067 PULMONARY FUNCTION EVALUATION OF SCHOOL CHILDREN BORN WITH VERY LOW BIRTH WEIGHT WITH AND WITHOUT BRONCHOPULMONARY DYSPLASIA

Emília da Silva Gonçalves; Francisco Mezzacappa Filho; Fernando Augusto de Lima Marson; André Moreno Morcillo; Adyélia Aparecida Dalbo Contrera Toro; José Dirceu Ribeiro. UNICAMP

Objective: Assess pulmonary flows of school children who were born with very low birth weight (VLBW) with and without bronchopulmonary dysplasia (BPD). Material: A total of 54 school children who were VLBW infants. Methodology: Observational and cross-sectional study. Asthma was diagnosed using the International Study of Asthma and Allergies in Childhood (ISAAC) which was answered by the parents and the school children underwent clinical examination and spirometry. The following variables were analyzed: asthma, bronchopulmonary dysplasia, age, birth weight and spirometry. The spirometry analyzed forced vital capacity (FVC), forced expiratory volume in 1 sec (FEV₁), Tiffeneau ratio (FEV₁/FVC) and forced expiratory flow at 25–75% of FVC (FEF25–75%). The focus of this study was to evaluate lung function using spirometry. Results: Age at evaluation (BPD: 9.5 ± 0.85; no-BPD: 10.1 ± 0.86 years) and birth weight (BPD: 916.7 ± 251.2; no-BPD: 1171.3 ± 190.5) were lower in the BPD group (p < 0.05). The prevalence of asthma among VLBW infants was 17/54 (31.5%) and in the group with BPD 6/18 (33.3%). Of the 54 school children evaluated 43 met the criteria for spirometry. School children with asthma had lower FEV₂5–75%-scores (n = 16; -1.04 ± 1.19) compared to the group of patients without asthma (n = 27; -0.38 ± 0.93) (p = 0.049). There was no difference regarding the spirometry variables in relation to presence or absence of BPD. Conclusion: Pulmonary flows of small Airways were lower in children with asthmatic. There was no difference regarding the spirometry variables in relation to presence or absence of BPD.

068 MODIFIER GENES IN CYSTIC FIBROSIS ACTING ON BRONCHODILATORS RESPONSE

Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertuzzo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Maria Angela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Adyélia Aparecida Dalbo Contrera Toro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Antonídio Fernando Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas)

Objectives: To determine the inhaled bronchodilator response on CF by spirometry markers and to compare with modifier genes, with lung expression, related with inflammatory response. Material: Included 126CF patients with 2CFTR mutations identified (classes I, II and/or III). Methodology: Spirometry was performed based on international standards (European Respiratory Society). For spirometry were considered: FVC (forced vital capacity), FEV₁ (forced expiratory volume in one second of FVC), FEV₁/FVC and forced expiratory flow between 25–75 percent of FVC (FEF25–75%) after inhaled bronchodilator. 256 polymorphisms (pol) on 125 genes were considered for association study. For genetic analysis, OpenArrayTM platform (LifeTechnologies™) was used. SPSSv22.0 performed the statistical analysis considering the percentage of bronchodilator response. Results: Data distribution was: (i) FVC: 1.56 ± 8.35; median = 1 (-20 to 32); (ii) FEV₁: 3.39 ± 7.70; median = 3(-1.2 to 48); (iii) FEV₁/FVC: 2.55 ± 7.57; median = 2(-19 to 33); (iv) FEF25–75%: 12.01 ± 26.48; median = 11.50(5.51 to 117). For spirometry markers and modifier genes, we have: (i) FVC%: was associated with 10 genes[10 pol](IL1B, IRAK1, IRAKM, NOS3, SELL, TBX21, TLR1 and TNFSF13b]; (ii) FEV1%: was
associated with 12 genes(13 pol) [ALOX5AP, IL18R1, IRAK3, ITGB5, NR3C1, ORMDL3, SELL, TBX21 and TSLP]; (ii) FEV1/FVC: was associated with 4 genes(5 pol)[ADCYAP1R1, CA10, IRF3, ITGB5, NR3C1, RUNX1(2pol), TLR1 and TOP2A]; (iv) FEF25-75%: was associated with 7 genes(8pol)[CDH1, FCER, IL1R1, IRAK1, IRF5, ITGB5, NOD1, RUNX1, TBX21, TLR4 and TSLP]. Conclusion: The individualized bronchodilator response was observed. In the respiratory diseases, there is a scientific gap to explain the response, often antagonistic, to bronchodilators. We demonstrate that the response on CF is more complex than previously thought. Many genetic variants were associated with the bronchodilator response. The genetic profile is particular for each marker.

069 ASSESSED PHYSICAL ACTIVITY IN BRAZILIAN PATIENTS WITH CYSTIC FIBROSIS AND ITS ASSOCIATION WITH EXACERBATIONS AND LUNG FUNCTION

Monica Ninet Rodas Gonzalez (Hospital Universitário Professor Edgard Santos); Linley Nathalie Teixeira Rocha (Escola Bahiana de Medicina e Saúde Pública-Bahia-Brazil); Carla Hilario da Cunha D. (Pós-Graduação em Medicina e Saúde do Hospital Universitário Professor Edgard Santos); Carolina de Oliveira Augusto (Escola Bahiana de Medicina e Saúde Pública-Bahia-Brazil); Almério de Souza Machado-Júnior (Universidade Estadual da Bahia-Brazil); Centro de Referência em Fibrose Cística do Hospital Especializado; Regina Terse-Ramos (Departamento de Pediatria, Faculdade de Medicina da Bahia da Universidade Federal da Bahia-Brazil).

Objectives: Evaluate habitual physical activity (HPA) levels among adults with (CF) and explore the relationship, HPA, number of exacerbations and lung function. Material: Patients aged 18 years or older with a confirmed diagnosis, agreed to participate and clinically stable. Methods: Prospective design, methods, data management or decision to publish. Patients with CF (n=146) were recruited from the CF outpatient clinic at a tertiary hospital. HPA was measured as MET-minutes/week (-1) using the short-form International Physical Activity Questionnaire (IPAQ) that captures the time and number of days spent on vigorous, moderate and walking activities in the previous week. Lung function, height and weight recorded for both groups. Pulmonary impairment was classified as mild (FEV1 60–79% predicted) moderate (FEV1 41–59% predicted) or severe (FEV1 40% predicted), number of exacerbations was recorded. Continuous variables were summarized as means, sd and percentages were calculated for categorical variables, Spearman Correlation used between IPAQ physical activity and other continuous variables. Results: 47 patients, 61.7% women and 61% were not white skin color; mean(sd) age of 45.0(19) and mean(sd) FEV1 % predicted of 62.4(28.9). The mean(sd) reported level of walking activity was 631.9(1034.4), moderate activity was 1064(2497) and vigorous activities was 110.4(372.8). When the relationship between habitual physical activity vs lung function and HPA vs number of exacerbations were assessed regardless of gender, there were no significant associations. Females showed moderate correlation of HPA (moderate activity) with number of exacerbations (rs = -0.4, p = 0.04) and males showed moderate correlations between HPA (walking activity) with number of exacerbations (rs = -0.5; p = 0.03); with HPA (moderate activity) (rs = -0.5; p = 0.04). No correlation of HPA with lung function was found. Conclusion: patients mostly reported activity levels that ranged from walking to moderate activity, despite of lung function; association between HPA and exacerbations numbers was found.

071 LUNG FUNCTION ON CYSTIC FIBROSIS: WHAT EXPLAINS THE GENETIC?

Fernando Augusto de Lima Marson (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Carmen Silva Bertozzo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); MariaÂngela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Adhélia Aparecida Dalbo Contrera Toro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Antônio Fernando Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Direceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas).

Objectives: To determine the CF severity by spirometry markers and influences the amount of drug delivered to the patient. Financial supported: Novartis. The authors have no competing interests, and do not receive financial benefits. Novartis played no role in the design, methods, data management or decision to publish.

070 NEBULIZER SYSTEMS USED BY CYSTIC FIBROSIS PATIENTS FROM MINAS GERAIS (BRAZIL); ARE THEY WORKING PROPERLY?

Evanirso da Silva Aquino (AMAM – Associação Mineira de Assistência a Mucoviscidose, Instituto de Medicina Tropical – USP, Hospital Infantil João Paulo II); Alberto Andrade Vergara (Hospital Infantil João Paulo II); Gustavo Santana (AMAM – Associação Mineira de Assistência a Mucoviscidose); Luiz Vicente R. Ferreira da Silva (IMT-USP, Instituto de Criança – USP).

Objective: The aim of this study was to evaluate the performance of jet nebulizers used by Brazilian CF patients from the State of Minas Gerais, Brazil. Methods: 146 nebulizer systems including compressor (Pronel Ultra<sup>®</sup>) and jet nebulizers (Pari LC plus<sup>®</sup>) were brought by CF patients attending 3 CF Centres for review, and additional 15 new systems were used as controls. Compressor performance was evaluated by measures of operating pressures using the PARI PG 10<sup>®</sup> manometer. The total nebulized drug (TND) and dead volume (DV) were analyzed using differences of weight of each nebulizer before and after 10 min of nebulization of 2.5 ml of saline. A questionnaire concerning usage and maintenance of the system was applied. Statistical analysis employed R package version 2.15, and alpha adopted was 0.05. Results: A significant proportion of compressors (39%) generated inadequate pressures, with values of less than half of new systems (controls), but working in the normal range. A wide range of TND and DV results was observed, with a significant correlation between TND & operation pressures (p < 0.001), and a negative correlation among DV & operation pressures (p < 0.001). These correlations were also observed in the controls, but with less dispersion of values. Significant lower values of TND and DV were observed among nebulizers working with inadequate pressures. Most patients report inadequate maintenance and infrequent substitution of parts. Conclusions: A significant proportion of nebulizer systems of CF patients from Minas Gerais is not working properly and maintenance is inadequate, a scenario that is probably occurring in the whole country. The pressure generated seems to be a critical aspect of system’s function and influences the amount of drug delivered to the patient. Financial supported: Novartis. The authors have no competing interests, and do not receive financial benefits. Novartis played no role in the design, methods, data management or decision to publish.

Neubulizer systems used by cystic fibrosis patients from Minas Gerais (Brazil); are they working properly?

Evanirso da Silva Aquino (AMAM – Associação Mineira de Assistência a Mucoviscidose, Instituto de Medicina Tropical – USP, Hospital Infantil João Paulo II); Alberto Andrade Vergara (Hospital Infantil João Paulo II); Gustavo Santana (AMAM – Associação Mineira de Assistência a Mucoviscidose); Luiz Vicente R. Ferreira da Silva (IMT-USP, Instituto de Criança – USP).

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S28 Abstract

II.R1 (2 pol), IL23R, IL33, ILIR2, NOS3 and RUNX1 (2 pol); (iii) FVC%: association with ten genes (10 pol) [CD40LG, HRH4, IL1B, IL3, ILIR2, IRF5, LRRC43, LTA4H, SELL and WISP]; (iii) FEV1: association with 12 genes (13 pol) [CD40LG, CTNNB1, FcerIA, HRH4 (2 pol), IL1B, IL3, IL6R, IRF5, NR3C1, SELL and WISP]; (iv) FEV1/FVC: association with 4 genes (5 pol) [BIRC5, IL23R (2 pol), IL2RB and IL4]; (v) FEV25-75%: 7 genes (8 pol) [BIRC5, HRH4 (2 pol), IL1B, IL6R, IRF5, NR3C1 and SMAD3]. Conclusion: Multiple genetic variants were associated with lung function on CF. The markers of lung disease — SaO2 and spirometry — have genes in common, but in many cases, we observed genetic peculiarities that characterize the individual severity of CF.

072 CLINICAL COURSE OF ASTHMA IN CHILDREN TREATED WITH OMALIZUMAB: A MULTICENTER STUDY IN COLOMBIA

Olga Lucia Morales Medina 1 (Universidad de Antioquia y Hospital San Vicente Fundacion); Laura Fernanda Niño Serna 2 (Hospital Pablo Tobón Uribe); Ángela María Pedraza 3 (Universidad el Rosario) 1 Pediatric Pulmonologist, Department of Pediatrics, University of Antioquia, San Vicente Foundation Hospital, Medellin, Colombia. 2 Pediatrician, Department of Pediatrics, Pablo Tobón Uribe Hospital, Medellin, Colombia. 3 Pediatric Pulmonologist, Military University, El Rosario University, San Rafael Clinical Hospital, Bogotá, Colombia.

Objective: To describe the clinical characteristics of pediatric patients aged 6 to 17 years diagnosed with moderate to severe allergic asthma undergoing treatment with omalizumab (OM) from January 2007 to February 2014 in 6 cities in Colombia. Methods: This retrospective, longitudinal cohort study included 63 children and adolescents. Evaluated outcomes included symptoms control, exacerbations frequency, decreased use of asthma control drugs and presence of adverse events before starting treatment and after 4 and 12 months of OM use. Results: After 4 and 12 months of treatment 89% and 96.6% of children under 12 years of age (p = 0.0015) and 78% and 91.6% of children over 12 years of age (p = 0.0145) had good symptom control, respectively. Furthermore, after 12 months, the number of exacerbations decreased to 2 (p = 0.00), the frequency of beta2 rescue inhaler use decreased to 0.37 days (p = 0.00), and the use of controller drugs decreased in 73% of patients, including decreased use of inhaled corticosteroids in 62%. Only one severe adverse event was reported in one patient. Conclusion: OM is an effective drug for the clinical control of moderate and severe allergic asthma. It reduces asthma symptoms and exacerbations and exhibits good tolerance and few severe adverse events.

073 IMPACT OF THE PROVISION OF FREE ASTHMA MEDICATIONS ON HOSPITAL ADMISSIONS FOR ASTHMA IN BRAZIL

Victoria daCrescêncio Silveira; Talitha Comaru; Frederico Orlando Friedrich; Paulo Márcio Pitres; Leonardo Araújo Pinto; Department of Pediatrics, Centro Infantil, Institute of Biomedical Research, PUCRS; Porto Alegre, Brazil.

Objective: since June 2011, the Brazilian health system provided asthma medicines completely free of charge to the patients with asthma. The aim of this study was to evaluate the impact of the provision of free asthma medications on hospital admissions for asthma in Brazil, using the national hospitalization database, and comparing the incidence of admission before and after the supply of these drugs. Non-respiratory admissions were used as comparison during the same time frame. Posteriorly, we analyzed the cost-effectiveness of this measure. Material and Methods: admissions of patients with 1 to 49 years of age by the Brazilian public health system with the diagnosis of asthma was compared pre (2008–2010) and post (2012–2014) provision of free medicines. Number of hospital admissions due to asthma and non-respiratory diseases were obtained from DATASUS, the Brazilian government open-access public health database system. The amount spent with asthma hospitalization was also obtained from DATASUS. Two main groups were selected: children and teenagers aged from 1 to 19 years and adults from 20 to 49 years old. Results: Admission rates for asthma strongly decreased from 90.09/100.000 (2008–2010) to 59.85/100.000 (2011–2014), when the period pre and post provision of free medicines were compared. These results suggest a significant reduction of the total hospitalization for respiratory diseases [OR 0.67 (CI 0.48 to 0.92)]. Non respiratory admission rates remained stable, when both periods were compared. In terms of economy, there was a decrease of 31% to the public expenditure (63 million in 2008 to 43 million in 2014). Conclusion: Asthma hospitalization rates decreased significantly in the three-year period after the provision of free medicines to treat asthma, whereas the rates of non-respiratory admissions remained stable during the same period. The savings to the public income were major and relevant.

074 REPRODUCIBILITY OF SPIROMETRIC MANEUVERS IN HEALTHY SCHOOL CHILDREN

Rafaela Coelho Minsky (Universidade do Estado de Santa Catarina); Francielli Camila Mucha (Universidade do Estado de Santa Catarina); Patrícia Morgana Rentz; Keil (Universidade do Estado de Santa Catarina); Tatiana Godoy Bobbio (Colaboradora do Nufipp); Camila Isabel Santos Schivinski (Universidade do Estado de Santa Catarina)

Aim: Evaluate the reproducibility of forced spirometric maneuvers during the examination by healthy school children. Methods: Observational, transverse analytic research with healthy children between 6 and 12 years. To confirm the healthiness of children were applied questionnaires and spirometry maneuvers (Pneumatógrafo Master Scope IOS/Jaeger®; Germany) according to American Thoracic Society, with FVC and FEV1 > 80% predicted. Children were separated into 3 groups: A) children who were able to perform spirometry maneuvers forced after 3 attempts; B) who performed in 4 attempts and C) those requiring between 5 and 8. Applied the Kolmogorov-Smirnov normality test, then the paired t test or Wilcoxon for comparison of spirometric measurements in the first and last maneuver performed by individuals of each group. Reproducibility of the maneuvers was determined by intraclass correlation coefficient of two-way (ICC) and used the Bland and Altman graphic. Significance level was 5% (p < 0.05). Results: Participated 149 children with a mean age of 9.22 ± 1.91. A group consists of 58 children, 35 girls, whose comparison of performance between the first and last spirometric maneuver showed significant increase to the absolute values of FVC and FEV1 (p < 0.05), with ICC = 0.96 and 0.98, respectively. In the analysis of 55 children in B group (32 male), there was an increase (p < 0.002) of the absolute values of the variables FVC and PEF (ICC = 0.96 and 0.84 respectively) as well as the predicted values (ICC = 0.82 and 0.32). Comparing the first and last maneuver of participants in the C group (36 children, 21 girls), only predicted values (ICC = 0.41) and absolute (ICC = 0.30) of PEF increased (p < 0.02). Conclusions: Reproducibility of forced spirometric maneuvers during testing performed by healthy schoolis dependent up on the number of attempts and the parameters analyzed, whether absolute or percentage of predicted. Only the percentage of predicted FEV1 was reproducible, respecting the number of 8 attempts.

075 DEMOGRAPHIC DATA AND SLEEP COMPLAINTS AS RISK FACTORS FOR OBSTRUCTIVE SLEEP APNEA IN CHILDREN – A REVIEW OF 19 YEARS IN A SLEEP CENTER

Cristiane Fumo dos Santos (Associação Fundo de Incentivo à Pesquisa, Universidade Federal de São Paulo); Gustavo Antonio Moreira (Associação Fundo de Incentivo à Pesquisa, Universidade Federal de São Paulo); Marcia Pradella-Hallinan (Universidade Federal de São Paulo); Beatriz

Pediatric Pulmonology
Introduction: Obstructive Sleep Apnea (OSA) is frequent in childhood. The gold-standard evaluation is polysomnography but it is a costly and not always available method. Objectives: Investigate data from history that would help in the diagnostic of OSA. Materials and methodology: Retrospective, cross-sectional study. Inclusion criteria $\geq 7$ and $< 18$ years, polysomnographic (PSG) evaluation between April 1995 and November 2014 at Instituto do Sono, São Paulo, Brazil. Exclusion criteria: cerebral palsy, neurodevelopmental delay, and genetic syndromes. An ordered logistic regression with hierarchical approach (first step, demographic data; second step sleep complaints) was used with intensity of OSA as outcome (not, mild, moderate, and severe). Results: We evaluated 2885 patients, mean age $12.6 \pm 3.2$ years, 62.8% male. An adjusted analysis for age, sex, family income, parent’s education, nutritional status, frequency and intensity of snore, difficult breathing, oral respiration, and enuresis was performed. Adjusted analysis showed that a higher intensity of OSA was associated with age (OR 1.05, IC 95% 1.03–1.08), male sex (OR 1.79, IC 95% 1.52–2.11), $< 2$ minimum wage (OR 1.27, IC 95% 1.19–1.35), parent’s education $\leq 4$ years (OR 1.71, IC 95% 1.08–2.70), BMI z-score (OR 1.27, IC 95% 1.19–1.35), snore $> 3$ times/week (OR 1.52, IC 95% 1.07 – 2.14), intensity of snore (extremely high OR 5.80, IC 95% 2.87 – 11.73), and enuresis (OR 1.33, IC 95% 1.04 – 1.71). Conclusion: Age, male sex, poorer income, low parent’s education, higher BMI z-score, snore more than 3 times / week, higher intensity of snore, and enuresis are risk factors that should rise the suspicion of OSA in children.

**076 OXYGEN SATURATION, PERIODIC BREATHING AND APNEA DURING SLEEP IN INFANTS 1 TO 4 MONTHS OLD LIVING AT 3,200 METERS ABOVE SEA LEVEL.**

**Santiago Ucros** (Fundación Sante Fe De Bogota – Colombia); **Claudia Granados** (Universidad Javeriana – Bogota, Colombia); **Karem Purejo** (Clínica Shiao – Bogota – Colombia); **Fausto Ortega** (Hospital Luis Fernando Martínez, Cauca – Ecuador); **Fernando Guillen** (Hospital Del Río. Universidad Del Azuay, Cuenca – Ecuador); **Sonia Restrepo** (Hospital de La Misericordia – Bogota – Colombia)

Objective: To describe SpO2 Levels during sleep in infants aged between 1 and 4 months living at an altitude of 3,200 m. The secondary objectives were the description of periodic breathing (PB) and apnea indexes. Methods: This study was carried out in Cauca, an Ecuadorian city located at 3,200 m above sea level. Polysomnographies were performed in 18 healthy infants between 1 and 4 months old. Results: The median for PB was 87% and for PB 72%. The Central Apnea Index (CAI) showed a median of 30.5/hour. The CI median, in relation to PB was 19.9/hour while the median for isolated CAI was 5.4/hour. Conclusions: The SpO2 was significantly lower than what is reported at sea level, whilst and the PB and CAI were significantly different from what is reported at sea level. At this altitude what is reported at sea level, whilst and the PB and CAI were significantly different from what is reported at sea level.

**078 LIPID PROFILE DISORDERS OF CYSTIC FIBROSIS PATIENTS OF CHILDREN’S HOSPITAL OF BRASILIA**

**Ana Catarina Maragog Firmo de Araujo** (Residency Program – Hospital of Base do Distrito Federal); **Ana Cristina de Araújo Bezerra** (Hospital of Base do Distrito Federal and Hospital da Criança de Brasília José Alencar); **Michele Batista Spencer Holanda Arantes** (Hospital da Criança de Brasília José Alencar); **Luciana de Freitas Velloso Monte** (Hospital of Base do Distrito Federal, Hospital da Criança de Brasília José Alencar and Universidade Católica de Brasilia).

Objective: The objective of this study was to evaluate the lipid profile of patients with cystic fibrosis (CF) treated in the CF Center of Children’s Hospital of Brasilia. Methods: This is a retrospective study based on analysis of clinical and laboratory data obtained from the medical records of patients aged 2 to 19 years, in regular monitoring in the clinics of this hospital. Results: The study included 53 CF patients. Hypertriglyceridemia was detected in 16 (30.2%) patients and hypercholesterolemia in 13 (24.5%). Five (9.4%) patients had higher than desirable LDL and 27 (50.9%) had HDL cholesterol below 45 mg/dL. Four (7.5%) patients had isolated hypercholesterolemia, 6 (11.3%) had isolated hypercholesterolemia and 14 (26.4%) had low HDL alone. In this study, the lipid profile of patients was not associated with age, body mass index, pancreatic function, physical activity, use of corticosteroids, FEV1 or Shwachman score. Conclusion. Lipid disorders were present in the CF patients of our study, with a predominance of hypertriglyceridemia and low HDL value. Due to increased life expectancy of individuals with CF, it is critical to conduct interventions at an early age with the objective of maintaining the health of these patients.
prospective studies aiming to answer what would be the causes and consequences of dyslipidemia in this specific group.

079 TUBERCULOSIS CONTACT TRACING IN CHILDREN BASED ON SIGNS AND SYMPTOMS

Paula da Nascimento Maia; Clemax Couto Sant’Anna; Maria de Fátima Bazuní Pombo March; Paola Andrea Vaca González; Bruna de Paula Santana; Sidnei Ferreira.
Instituto de Puercicultura e Pediatria Matogrossense Governor UFRJ.

Objective: Evaluate tuberculosis (TB) contacts based on signs and symptoms, according to Brazilian Guidelines. Material and Methodology: Retrospective cross-sectional study of children and adolescents who were TB contacts in a health center of Rio de Janeiro, Brazil, between 2006 and 2014. They all were evaluated by: tuberculin skin test (TST), chest X-ray and presence and duration of symptoms. Subsequently the contacts were classified as active TB, latent infection of tuberculosis (LTBI) and exposed. Results: 245 contacts were assessed, of which 236 were included in the study. Of these, 181 (76.7%) were asymptomatic, 30 (12.7%) had symptoms by less than 2 weeks and 25 (10.6%) had symptoms by 2 or more weeks. Among asymptomatic contacts, 125 (69%) had LTBI, 3 (1.6%) had active TB and 39 (21.6%) were exposed. Among contacts with symptoms by less than 2 weeks, 16 (53.3%) had LTBI, 5 (16.7%) had active TB and 7 (23.3%) were exposed. Among symptomatic patients by 2 or more weeks, 13 (52%) had LTBI, 4 (16%) had active TB and 6 (24%) were exposed. Conclusion: In Brazil, TB contact tracing in children based on signs and symptoms should be complemented by TST and chest X-ray, allowing better identify active TB and LTBI. Most asymptomatic patients had LTBI but 1.6% had TB disease. Among symptomatics (regardless of duration), 16.7% had active TB.

080 FREQUENCY OF THE PHE508DEL MUTATION AND ITS CLINICAL CORRELATIONS IN PATIENTS WITH CYSTIC FIBROSIS IN A HIGHLY ADMIXED POPULATION IN NORTHEASTERN BRAZIL

Edna Luíza Santos de Souza (School of Medicine of Bahia, Federal University of Bahia); Luís Ribeiro Mota (Institute of Biology - Laboratory of Human Genetics and Mutagenesis Federal University of Bahia); Carlos Sidney Silva Pimentel (School of Medicine of Bahia, Federal University of Bahia); Paloma Hoarej Bittencourt (School of Medicine of Bahia, Federal University of Bahia); Salmo Raskin (Pontificia Universidade Católica do Paraná (PUCPR); Renata Luíza Leite Ferreira de Lima (Institute of Biology - Laboratory of Human Genetics and Mutagenesis Federal University of Bahia).

Objectives: To determine the prevalence of the Phe508del mutation in patients receiving care at the multidisciplinary cystic fibrosis (CF) outpatient department in a Teaching Hospital and to investigate the association between the presence of the Phe508del mutation and the clinical manifestations presented by the patients. Material and Methods: This was a cross-sectional study. CF patients under the age of 21 years with CF followed up at outpatient clinics in a Teaching Hospital between March 2005 and August 2015 were included. A questionnaire was completed to obtain clinical and epidemiological data, followed by blood sample collection from the patients for molecular analysis of Phe508 mutation. Descriptive analysis was performed for all variables. The gene frequencies were determined by dividing the number of mutant alleles by the total number of alleles in the sample. Results: Fifty three unrelated patients were included, 50.9% were female. All the children were classified as non-white. The mean age of the patients at admission to the study was 10.4 years. The mutation was detected in 33.9% (18/53) of the patients, with an allele frequency of 25.5% (27/106). Fifty percent of the patients were homozygous (9/18). The median age of the patients was 5.6 years at the time of diagnosis. The earliest onset of symptoms was the first month of life, with symptoms beginning this early in 15 children (28.3%). Around forty percent of the patients affected by the Phe508del mutation had the onset of symptoms in their first month of life. Pancreatic insufficiency (IP) was recorded in 44 patients. All but one patient with Phe508del mutation had IP. Conclusions: The frequency of the Phe508del mutation was lower than the one found in Caucasian populations, thus reflecting the racial/ethnic characteristics of the analyzed population. Diagnosis of the disease was late and symptoms were more severe and occurred earlier in patients with the Phe508del mutation.

081 ASSOCIATION OF NUTRITIONAL AND SPIROMETRIC EVALUATION OF CHILDREN WITH CYSTIC FIBROSIS

Mirella Aparecida Neves Almeida (Hospital Infantil Pequeno Príncipe); Jocemara Guirmoni (Hospital Infantil Pequeno Príncipe); Denise Tiemi Miyakawa (Hospital Infantil Pequeno Príncipe); Dylanne Marcia Lopes Bastos (Hospital Infantil Pequeno Príncipe); Gedson Luiz Picharski (Instituto De Pesquisa Péel Pequeno Príncipe).

In cystic fibrosis (CF), the nutritional deficiency is a result of pancreatic insufficiency, malabsorption and obstructive lung disease. The disease often manifests as poor growth and weight gain during childhood and previous studies have shown that worsening pulmonary function is associated with undernutrition. A multidisciplinary therapy not only targets in lung disease but also focuses on gastrointestinal manifestations and nutritional deficiency. The clinical evaluation of measures of growth and body composition provide interventional strategies when patients are at risk for undernutrition or with nutritional failure. The purpose of this study was to evaluate the relationship between the nutritional status and pulmonary function in children with Cystic Fibrosis. Retrospective study involving of 25 patients (6 - 18 years of age) under clinical follow up treatment with a Multidisciplinary Cystic Fibrosis Outpatient Clinic at The Children’s Hospital Pequeno Príncipe in the city of Curitiba-PR-Brazil. Information about nutritional assessment, body composition and pulmonary function was evaluated. We studied 25 patients who where able to perform spirometry (13 male and 12 female), 20 (80%) with pancreatic insufficiency, 22 (88%) identified after 1 year of age, 19 (76%) with airflow limitation moderate and severe (.... Classification) and 3 with Cystic Fibrosis Related Diabetes (CFRD), 73.7% patients were with airflow limitation moderate or severe and all of them had pancreatic insufficiency. Amongst patients with z score of BMI for age<-2, 68% are with airflow limitation. The worst pulmonary function (FEV1 = 44%) were associated with the smaller tricipital skinfold measurements. Even those with adequate fat free mass (FFM), 14 (56%) were with low FEV1 values.

The present data demonstrate that in Cystic Fibrosis there are more factors that influence the pulmonary function, not only the nutritional status and body composition.

082 DOSIMETRIC EVALUATION OF RADIOGRAPHIC EXAMS PERFORMED IN PREMATURE CHILDREN DURING THE HOSPITALIZATION PERIOD IN A NEONATAL INTENSIVE CARE UNIT

Marcia Larissa Cavallari da Costa Gois (Faculdades Pequeno Príncipe); Hugo Reuters Schelin (Faculdades Pequeno Príncipe); Valeriy Denyak (Faculdades Pequeno Príncipe); Jorge Ledesma (Hospital Pequeno Príncipe); Adriano Legnani (Hospital Pequeno Príncipe); Ana Paula Bunick (Faculdades Pequeno Príncipe).

Objective: Evaluate radiographic exams of premature children in a neonatal intensive care unit (NICU) during the period of hospitalization. Material and Methodology: In this study were included all premature hospitalized in a NICU between January 2013 and December 2014, with a birth weight of less
than 1500g. Full-term patients with birth weight greater than 1500g and patients who died during the hospitalization period were excluded from the analysis. Data collection occurred in 3 phases. In the first and second phases, data were collected from medical records of the patients and from technical parameters documented in the radiographers’ record book, respectively. In the third phase, the ionizing radiation dose assessment received during the exams was verified. Results: In this study were included 57 premature children with a median birth weight of 1128g and a median gestational age of 29 weeks. The average value of permanence of NICU stay was 73 days. The hospital has 17 radiographers that together performed 1128 radiographs, giving an average number of 19.7 radiographs per patient. A significant difference is observed in the dispersion of the technical parameter kVp used by one of the radiographers. The difference in median kVp values reaches up to 20% and it might cause a dose-doubling of the received ionizing radiation. The variety of the technical parameter mAs was different between radiographers 10 and 17 and between radiographers 4 and 16, which might lead to dose tripling. In most cases the premature children received a dose rate less than 28 Gy/day, with the largest number of infants receiving a dose of ~10 Gy per day. Conclusion: It is necessary a large amount of radiographic examinations for the diagnosis and treatment of premature children during their hospitalization period in a NICU. The obtained dose rate shows that the standardization of the technique used by the radiographers could reduce the radiograph dose by 20 – 30 Gy per examination.

083 EVALUATION OF CARBON DIOXIDE PRODUCTION DURING SUBMAXIMAL TREADMILL STRESS TESTS IN CYSTIC FIBROSIS PATIENTS

Paloma Lopes Francisco Parazzi (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Maria Angela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Camila Isabel Santos Schivinski (State University of Santa Catarina, Center of Physical Education and Sports); Antonio Fernando Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Direceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas)

Objectives: With the progressive deterioration of the pulmonary disease, the increase in dead space demands changes in pulmonary ventilation to maintain adequate alveolar ventilation during exercise. Therefore, changes in lung function throughout time are correlated to changes in exercise capacity. The increase in blood CO₂ can rapidly result in a respiratory acidosis; therefore, the production of carbon dioxide (VCO₂) is one of the primary factors to determine the pattern of ventilation during exercise. In this context, our objective was to evaluate and compare VCO₂ at rest, during and after submaximal exercise in children, adolescents and young adults between six and 25 years of age, with cystic fibrosis (CFG) and without cystic fibrosis (CG). Material: A controlled, clinical trial, with 128 individuals, 64 with cystic fibrosis (CF), of both genders, from a University Hospital. Methodology: All individuals completed the submaximal stress test on the treadmill for six minutes and other exams, after the approval of the Institutional Ethics Committee and signed Informed Consent form. The comparison between the groups of patients with CF and the control group was made through Kruskal-Wallis and Mann-Whitney tests. Results: The values of VEF1 and VCO₂ showed a direct association in all time points analyzed (p<0.001). Conclusion: The retention of VCO₂ by the CFG during the submaximal stress test can be explained due to the specific changes in gas exchange present in this pulmonary disease: disorganization of the lung architecture, destruction of parts of the capillary bed, increase in dead space and secretion retention.

084 EPISODIC SEASONAL PSEUDO-BARTTER SYNDROME IN CYSTIC FIBROSIS: CASES REVIEW

Patrícia Fernandes Barreto Machado Costa; Natália Kopke Soares Nascimento; Tania Wrobel Folesca; Deborah Aragão de Poio Silveira; Renata Wrobel Folesca Cohen; Renato Farm D’Amodio; Instituto Nacional da Saúde da Mulher, Criança e Adolescente Fernandes Figueira-FIOCRUZ

Objective: To describe clinical and laboratorial data from infants with Cystic Fibrosis (CF) diagnosis and clinical presentation of Pseudo-Bartter syndrome (metabolic alkalosis, hypochloremia and hyponatremia). Material: Infants with CF diagnosis and clinical presentation of pseudo-Bartter syndrome (PBS) who were followed at this CF reference center. Methods: After approval from ethics committee, retrospective data were collected from medical reports. Results: From January 2013 to September 2015, 43 infants had CF diagnosis. 25/43 came from neonatal screening program. Of these 7 (16,7%) had Pseudo Bartter syndrome and 2 presented recurrence of PBS (total of 9 episodes). 6/7 were female, and mean age at CF diagnosis was 4 months (3-6 months) while mean age at pseudo-Bartter episode was 6.5 months. This episode happened during summer for 6/9 events and on spring for 3/9. Three infants (3/7) presented only metabolic disease, while other 4/7 had also respiratory exacerbation and 2/7 had malnutrition criteria. All patients had underactivity, weight loss and dehydration signs. Facial/ extremities edema was found in 6/7 infants and hepatomegaly in 6/7. Blood biochemistry revealed mean sodium, potassium and chloride dosage of 127 (126–131), 2,31(1,4–3,0) e 84,7 (68–110) mEq/L, respectively. All infants had metabolic alkalosis with mean pH of 7.59 (7.52–7.65) e bicarbonate of 43 mmol/L (35–56,1). Renal function was preserved, with mean values for urea: 46 mg/dl (31–96), creatinine: 0,52 mg/dl (0,32–0,77). All patients were admitted to hospital. Conclusion: Neonatal screening contributed for early diagnosis and treatment of CF infants with metabolic disease, helping differential diagnosis of Bartter syndrome. We highlight that CF must be considered as a possible diagnosis for infants with this metabolic syndrome.

085 VASOOCCLUSIVE CRISIS AND SLEEP QUALITY IN CHILDREN AND ADOLESCENTS WITH SICKLE CELL DISEASE

Tatiane Anunciação Ferreira (Curso de Pós-Graduação em Medicina e Saúde Humana); Sálvian Henrique de Souza Miranda (Faculdade de Medicina da Bahia – Universidade Federal da Bahia-Brazil); Renata Maria Pereira Vieira Barbosa (Faculdade de Medicina da Bahia – Universidade Federal da Bahia-Brazil); José Cleomarque Leite Júnior (Faculdade de Medicina da Bahia – Universidade Federal da Bahia-Brazil); Luísa Danielle Alves de Souza Santos (Faculdade de Medicina da Bahia – Universidade Federal da Bahia-Brazil); Regina Terse-Ramos (Departamento de Pediatra – Faculdade de Medicina da Bahia – Universidade Federal da Bahia-Brazil)

Aim: to verify the relationship between vasoocclusive crisis and sleep quality in children with sickle cell disease (SCD). Material: The study involved 59 SCD and 62 healthy patients aged 6–18 years. Sleep disturbance was evaluated using the Sleep Disturbance Scale for Children (SDSC) which assesses five subdomains: disorders of initiating and maintaining sleep (DIMS), sleep breathing disorders (SBD), disorders of arousal (DA), sleep-wake transition disorders (SWTD), disorders of excessive somnolence (DOES), and sleep hyperhidrosis (SHY). Methods: Cross-sectional study; patients were recruited from the SCD outpatient clinic. Number of vasoocclusive crises (VOC) was recorded and subsequently categorized as ≤ 2 and > 2. Means and standard deviations and percentages were calculated for categorical variables; the Mann–Whitney U test, Spearman Correlation and p < 0.05 were used. Results: Mean(sd) age of case and comparison groups
were 12.6 ± 3.1 vs. 11.6 ± 3.0; p = 0.10, respectively; most were males in the SCD group (p = 0.01). The means(±) of the overall score of the SDSC scale between the case and comparison groups were 47.7 ± 11.5 vs. 45.6 ± 12.7; p = 0.8; in the specific subscales, DOES and SBD prevailed in the SCD group: 7.1 ± 3.2 vs. 9.1 ± 3.2; p = 0.02; and 6.7 ± 3.4 vs. 5.2 ± 2.7, p = 0.04 respectively. When the median values of sleep disorders domains were compared respectively to groups with ? 2 and ? 3 VOC, statistical significance was only found in DA subdomain (p = 0.01). It’s found significant correlation between number of CVO with DA (rs = 0.3 p = 0.02) and overall score of the SDSC scale (r = 0.3 p = 0.02), CONCLUSION: The case group had higher values of DOES and SBD comparing to the healthy group. The case group with more exacerbations presented more DA and there was a significant correlation between number of VOC with DA and overall score of the SDSC scale.

086 STENOTROPHOMONAS MALTOPHILIA: PREVALENCE AND CLINICAL SIGNIFICANCE IN 12 YEARS OF FOLLOW-UP IN HC-UNICAMP

Mauro Pedromonico Arrym; Talita Bianchi Aiello; Renan Marrichi Mauch; José Dirceu Ribeiro; Antonio Fernando Ribeiro; Carlos Emilio Levy; HC-Unicamp

Introduction: Stenotrophomonas maltophilia is an opportunistic bacteria and potential pathogen in patients with cystic fibrosis (CF). Although the recent increase in the prevalence of infection with S. maltophilia in this population, its importance in the progression of lung disease is not evident yet. Objective: Evaluate the prevalence of S. maltophilia in CF patients treated at the referral center of the HC-Unicamp, from 2002 to 2014. Method: A review of the microbiological positive cultures from respiratory sample with S. maltophilia from Microbiology Laboratory database. Results: A total of 69 patients showed 155 positive samples during the period of study and 37 (53.6%) were male. Of these, 52 (75.3%) with ? 2 positive samples and 17 (24.7%) with ? 3 positive samples. We observed an increased number of patients with isolation of S. maltophilia in the last two years of follow-up, corresponding to 33% of the total samples. In 12 years, only 3 patients had chronic infection, on average of 33 years old (range 8–41). Conclusion: The prevalence of S. maltophilia is relevant, considering that our center attending 250 patients, but their clinical significance seems more restricted to adult patients with chronic lung disease and associated with other opportunistic agents. However, the recent increased in the isolation of this pathogen must be followed to assess their role in the transient colonization and their true clinical significance.

087 WHAT THE GENETIC “SPEAKS” ABOUT THE ALLERGIC ASTHMA

Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertazzo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Andressa de Oliveira Peixoto (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Maria Angela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Tânia Kawasaki de Araújo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); José Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas)

Objectives: To determine the genetic profile of allergic asthma (AA) in a Brazilian population, considering genes related to inflammatory responses. Material: Included 221 AA patients, aged between 6–18 years, and 536 healthy controls aged over than 18 years with no personal and/or family history of lung illness in 2 generations. Methodology: 256 polymorphisms (pol) on 125 genes were considered for association study. For genetic analysis, OpenArrayTM platform (LifeTechnologies®) was used. SPSS®v22.0 performed a Pearson correlation analysis. Result: Comparing the AA and healthy control subjects there was difference for 56 genes, corresponding to the association with 86 pol. In the sample, 79 genotypes were associated with increased risk for AA and 80 genotypes as protection factor for the disease. Odds ratio (OR), as risk factor for AA, ranged from 6.4 (95% CI:1.01–10.2) for the rs35904768 AC pol in TBX2AR gene to 1.43 (95% CI:1.01–2.01) for the rs11071559 CC pol in RORA gene. Regarding the protective factor for AA, OR ranged from 0.7 (95% CI:0.05–0.099) for the rs2228145 AC in IL6R gene to 0.10 (95% CI:0.01–0.71 for the rs971998 TT pol in IL18RAP gene and 0.10 (95% CI:0.04–0.23) for rs63344 GG pol in NTRK1 gene. In the sample, there was association of 56 pol in the heterozygous genotype (over-dominance) for the AA – demonstrating a genetic heritage character of keeping of the asthma with high prevalence. Conclusion: Our results confirm the genetic variability that determines AA in our population. Multiple genes form an intrinsic network of associations that lead to the presence of complex disease that needs to be well studied to be understood from a genetic point of view. Possibly, we have for each individual “an” AA, with a single genetic basis and environment conferring the disease.

088 OXACILLIN-RESISTANT STAPHYLOCOCCUS AUREUS: AN EMERGING PATHOGEN IN OUR SERVICE?

Talita Bianchi Aiello; Mauro Pedromonico Arrym; Renan Marrichi Mauch; José Dirceu Ribeiro; Antonio Fernando Ribeiro; Carlos Emilio Levy; HC-Unicamp

Introduction: Staphylococcus aureus is one of the first potentially pathogenic bacteria detected in cystic fibrosis (CF) infants and children. The rise of oxacillin-resistant S. aureus (ORSA) in recent years can be reflection of the frequent and prolonged use of antimicrobials but its clinical significance in CF is still not clear. Objective: To determine the prevalence of oxacillin-resistant S. aureus in CF patients attending referral center of the HC-Unicamp, from 2002 to 2014. Method: A review of the microbiological positive cultures from respiratory sample from the Microbiology Laboratory database. Microbial sensitivity tests were performed according to the CLSI recommendations. Results: An approximate total of 250 patients, 60 showed 212 ORSA samples during the period and 33 (55%) were male. Of these, 36 (60%) with ? 2 positive samples and 24 (40%) with ? 3 positive samples. It stands out the considerable increased in the isolation of this pathogen in 2014, representing 22.2% of total positive samples over the 12 years of study. However, in this period, only 10 patients were classified as ORSA chronically infected, on average 15.5 years old (range 3–35) Conclusion: About 1/4 of our patients have been colonized by ORSA, but only 4% are chronic cases. Based on the recent increased number of positive samples, this pathogen in the last year, it’s worrying the possibility to change this profile, explaining greater monitoring of care and discussion of early intervention measures.

089 THE QUALITY OF THE SWEAT TEST CONSIDERING THE PROPORTION OF SODIUM AND CHLORIDE AS QUALITY PARAMETER: ARE WE ON THE RIGHT WAY?

Alethéia Guimarães Faria Guimarães Faria; Fernando Augusto de Lima Marson; Antônio Fernando Ribeiro; Carla Cristina Souza Gomez; Maria de Fátima Servidoni; José Dirceu Ribeiro; Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas

Objective: To verify the quality of the sweat test (ST) by the proportion of sodium (Na) and chloride (Cl) in the ST exam. Material: There was performed a retrospective analysis of 5,721 ST exams considering the values of Na and Cl. Methodology: The ST was considered with low quality in two conditions, if: (i) Cl < 60 mEq/L, Na should be greater than Cl; and (ii)
CI to 60 mEq/L, Na should be minor than Cl. Results: A total of 5721 ST were included. From all ST performed, two are not included considering the absence of the sodium values. Of the total of exams (5719), 5049 showed a correct proportion of Na and CI (88.3%) and 670 (11.7%) had limitations. The diagnosis result was divided into: (G) < 30 mEq/L (3670 – 64.2%); (GI) < 30 to < 40 mEq/L (655 – 11.5%); (GII) < 40 and < 60 mEq/L (673 – 11.8%); (GIV) > 60 mEq/L (721 to 12.6%). The number of tests with inadequate quality for diagnostic group were: GI – 190/3670 (5.2%) [OR: 0.179; 95%CI: 0.149 to 0.213]; (GII) 114/655 (17.4%) [OR: 1.709; 95% CI: 1.371 to 2.13]; (GIII) 202/673 (30%) [OR: 4.195; 95% CI: 3.468 to 5.075]; (GIV) 164/721 (22.7%) [OR: 2.614; 95% CI: 1.47 to 3.183] (p < 0.001). Conclusion: Considering that the quality of the ST depends on the proportion of sodium and chloride values, in our sample, we observe high number of ST with low quality, especially in GIII and GIV. However, we can consider the bigger frequency of limitations in GIII as an information that should be analyzed with caution. Maybe, in this group, the proportion between the ions could be a characteristic, and not a limitation.

090 EVOLUTION OF NUTRITIONAL STATUS IN PEDIATRIC CYSTIC FIBROSIS PATIENTS AT CENTER OF EXCELLENCE IN NORTHEASTERN BRAZIL: A COHORT STUDY

Karine Yoko Kodama Dantas; Ana Paula Brito de Aguiar; Carolina de Godoy Almeida; Tatiane da Anunciação Ferreira; Sandra Santos Valois; Edna Lucia Souza; Federal University of Bahia

Objectives: To assess the nutritional status of children with cystic fibrosis (CF) at the diagnosis and after one year of treatment at the Multidisciplinary outpatient department in a Teaching Hospital. Material and Methods: It was a retrospective cohort study. All the CF patients under the age of 18 years with CF confirmed by sweat test or genetic study followed up at outpatient clinics in a Teaching Hospital from June 2008 to June 2015 were included. The data were obtained from medical records. Nutritional status was classified on the basis of WHO BMI guidelines 2006 at time of diagnosis and one year after treatment. Descriptive analysis included calculations of means, medians, and standard deviation in the study variables. Results: Forty-six patients were included, 56.5% were male. The mean age at disease diagnosis was 77 months. At time of diagnosis, 24 (52.2%) children had some nutritional disorder. For patients under 2 years old malnutrition and short stature were more common (40%); severe malnutrition e severe short stature occurred in 30% and 70% of these patients, in that order. For children at 2 years or older, malnutrition and short stature were observed in 53.8% and 43.2% of patients, respectively. Thirty-eight patients were followed for one year after diagnosis and treatment and 13 (34.2%) continued to have some nutrition disorder: malnutrition and short stature (15.4%) for children under 2 years; malnutrition (23%) and short stature e (39%) for those older than 2 years. Conclusions: The frequency of nutritional disorders was high at the time of diagnosis, principally malnutrition and short stature for all age groups. Despite the treatment, around 40% of the followed patients remained malnourished and 50% continued to have short stature. This may be due to the late diagnosis of disease and the difficulties of treatment in the low-income populations.

091 CLINICAL SEVERITY OF CYSTIC FIBROSIS CONDITIONED BY rs1800482 (-954G>C) POLYMORPHISM IN THE NOS2 GENE

Stéphanie Villa-Nova Pereira (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertuzzo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Antonio Fernando Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Dircu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Fernando Augusto de Lima Marson (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas)

Objective: To associate the rs1800482 (-954G>C) polymorphism in the Nos2 gene (nitric oxide metabolism) with the cystic fibrosis (CF) severity. Material: Enrolled 177 CF patients. Methodology: The rs1800482 screening was done by the Restriction Fragments Length Polymorphism technique. To analysis association were used 43 clinical markers. The CF patients were grouped by CFTR mutations: (G) all patients; (GII) IM/IM; (GIII) IM/NIM; (GIV) NIM/NIM. IM, identified mutation; NIM, no identified mutation. All IM were class I, II or III. The rs1800482 was analyzed considering: complete genotype, GGVGCACCC, CCCsGCAGG and CCGsvsGCg. Conclusion: There was a trend for a higher Lund Mac Cay score as asymptomatic and 45,83% had nasossinusal complaints, mostly obstructive

092 NASOSSINUSAL COMPUTED TOMOGRAPHY CORRELATIONS IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS (CF)

Paula de Souza Dias Lopes; Michelle Mancini; Paulo José Cauduro Maróstica; Léa Sêkine; Otávio Bezjman Piltcher; Letícia Rocha Machado; Hospital de Clínicas de Porto Alegre

Objectives: To evaluate if there is a correlation between nasossinusal computed tomography (CT) findings and genotype, Pseudomonas aeruginosa (Pa) colonization, forced expiratory volume in 1 sec (FEV1), body mass index (BMI), symptoms, Schwachman-Kulczycki score (SK), age at the time of CT scanning was 9,16 years, at the time of the CT, followed at the Hospital de Clínicas de Porto Alegre pediatric CF center. CT findings were scored by Lund Mac Cay score. FEV1 and BMI were converted to z scores. Results: 62 patients were included, being 50% males. 30 had identified genotypes, 2 had at least one mutation. All IM were class I, II or III. The rs1800482 was analyzed considering: complete genotype, GGVGCACCC, CCCsGCAGG and CCGsvsGCg. Alpha = 0.05. Results: From 177 CF patients: CC – 5 (2.3%); CG – 40 (22.6%), GG 133 (75.1%). In the comparisons performed, positive association was find to: (i) complete genotype: IM/IM and FEV75% BD response (p = 0.046); IM/NIM and early digestive symptoms (p = 0.019), NIM/NIM and mucoid P. aeruginosa (p = 0.050); (ii) GGVGCACCC: (a) all patients: mucoid P. aeruginosa (p = 0.045); (b) IM/IM: FEV75% BD response (p = 0.046), Staphylococcus aureus (p = 0.032); (c) IM/NIM: diagnostic time (p = 0.027), early digestive symptoms (p = 0.014); (d) NIM/NIM: mucoid P. aeruginosa (p = 0.025); (iii) CCsGCAGG and all patients to BD response: FEV52% (p = 0.045), FEV50% (p = 0.023), FEV25-75% (p = 0.047) and FEV1/FVC (p = 0.014); (iv) CCGsvsGCg and IM/IM: FEV75% BD response (p = 0.046). Conclusion: The rs1800482 is a potential modifier gene of CF severity in our sample.

093 CLINICAL EVOLUTION OF INFANTS WITH CYSTIC FIBROSIS: WHAT HAVE WE LEARNED?

Caren Ishikawa; Rais André S. Watanebe; Marcela Duarte de Sillos; Beatriz N. Barbisan; Sonia Mayumi Chiba; Clovis Eduardo Tadeu Gomes; Universidade Federal de São Paulo

Pediatric Pulmonology
Objective: To describe the clinical evolution of infants directed to a reference center. Methods: To analyze medical records of infants with cystic fibrosis (CF) from February 2010 to October 2015. The minimum follow-up was three months. Main studied variables: presence of meconium ileus, age at diagnosis, beginning of the respiratory symptoms and pancreatic insufficiency (PI), nutritional evolution, causes of hospitalizations, medications, growth of Pseudomonas aeruginosa (Pa), Staphylococcus aureus (Sa) and Burkholderia cepacia (Bc) in the oropharyngeal cultures. Results: 37 patients were analyzed (6m0-5y), 33 of them were referred from the newborn screening (NS). The NS was negative in four cases. Twenty-one (57%) patients were male and nineteen (51%) were Caucasian. The median age at the diagnosis was 2 months (1–18 mo). Meconium ileus occurred in 11% of the patients and 86.4% presented PI. The medium age at the beginning of respiratory symptoms and PI were both of two months. In the comparison of Z-score of weight for age in the first and in the last consultation, was observed an improvement of this parameter in the majority of the cases. 97% of the patients needed hospitalization due to respiratory exacerbations, followed by hydroelectrolytic disturbances. Twenty-two patients were making use of dornase alfa and sixteen were using inhaled tobramycin in alternating months. Ten patients with hepatic disorder were receiving ursodeoxycholic acid. The oropharyngeal cultures demonstrated: 23 (62%) patients had a growth of Pa, 29 (78%) Sa (eight were oxacillin-resistant) and 4 (11%) had Bc. Conclusions: Nutritional recovery was satisfactory after the pancreatic enzyme replacement therapy and treatment of respiratory complications. Clinical manifestations were early, with a high rate of hospitalizations. There was isolation in the respiratory secretions of the common bacteria in CF. Even with an early diagnosis, the infants with CF had severe complications.

Objective: To describe clinical and laboratorial data (at the moment of diagnosis and one year later), of CF patients who came to this institution from neonatal screening test from January 2013 to September 2015. Material/Methods: Retrospective study based on analysis of clinical, laboratorial and therapeutic records at the moment of CF diagnosis and one year later. Results: 26 patients (14 males) had their CF diagnosis from neonatal screening test from January 2013 to September 2015. One year later, 29 (78%) of them were referred from the newborn screening (NS). The NS was negative in four cases. Twenty-one (57%) patients were male and nineteen (51%) were Caucasian. The median age at the diagnosis was 2 months (1–18 mo). Meconium ileus occurred in 11% of the patients and 86.4% presented PI. The medium age at the beginning of respiratory symptoms and PI were both of two months. In the comparison of Z-score of weight for age in the first and in the last consultation, was observed an improvement of this parameter in the majority of the cases. 97% of the patients needed hospitalization due to respiratory exacerbations, followed by hydroelectrolytic disturbances. Twenty-two patients were making use of dornase alfa and sixteen were using inhaled tobramycin in alternating months. Ten patients with hepatic disorder were receiving ursodeoxycholic acid. The oropharyngeal cultures demonstrated: 23 (62%) patients had a growth of Pa, 29 (78%) Sa (eight were oxacillin-resistant) and 4 (11%) had Bc. Conclusions: Nutritional recovery was satisfactory after the pancreatic enzyme replacement therapy and treatment of respiratory complications. Clinical manifestations were early, with a high rate of hospitalizations. There was isolation in the respiratory secretions of the common bacteria in CF. Even with an early diagnosis, the infants with CF had severe complications.

Objective: To assess the link between 11 viruses and the diagnosis of Cystic Fibrosis (CF). Materials and Methods: Patients (n = 49) with 2 IRT dosages changed (> 70 ng/ml) were referred for sweat test, according to the rules of Newborn Screening State of Sao Paulo. The saliva samples were collected with a roll-shaped cotton – Salivette® (Sarstedt-Germany – http://www.sarstedt.com) for three minutes, after fasting for 1 hr and embrocation with gauze moistened with deionized water. The samples were immediately centrifuged at 1500 rpm for 15 minutes, amounts less than 0.6 mL were discarded. Chloride concentration were analyzed by ABL gasometer (model 835, Radiometer®, Denmark) by gasometric analysis using an ion-selective electrode direct. was performed according to Gibson and Cooke. Results: Mean age in CF group, (GFC) = 1.25 years and in no CF group, (NCFG) 0.4 years. In CFG: Saliva chloride mean = 36.04 mmol/L; SD = 20.49 mmol/L and the standard error of the mean = 4.37 mmol/L and in Sweat chloride the mean = 118.6 mgEq/L; SD = 33.54 mgEq/L; standard error of the mean = 7.15 mgEq/L. In first IRT the mean = 223.41 mgEq/L; SD = 103.92 mgEq/L and median = 220.7 mgEq/L, in second IRT the mean = 201.55 mgEq/L, SD = 116.56 mgEq/L and median = 149.6 mgEq/mL. NCFG: Saliva chloride mean = 17.94 mmol/L; SD = 7.05 mmol/L and the standard error of the mean = 1.66 mmol/L; Sweat chloride the mean = 14.68 mgEq/L; SD = 5.95 mgEq/L; standard error of the mean = 1.40 mgEq/L. The values of IRT in this group was: First IRT mean = 80.77 ng/ml; SD = 31.77 ng/ml and median = 74.8 ng/ml; second IRT mean = 124.22 ng/ml; SD = 154.13 ng/ml and median = 84 ng/ml. Correlation Coefficient of Spearman of saliva chloride in versus first IRT = 0.36 (p = 0.27); saliva chloride versus second IRT = 0.59 (p = 0.000) and saliva chloride versus sweat chloride = 0.35 (p = 0.22). CONCLUSION: Therefore the IRT and the chloride concentration in the sweat, the chloride ion in the saliva was greater in individuals with CF, which suggests the possibility to use the dosage chloride concentration in saliva for both screening and for diagnosis of CF.

Objective: To assess the link between 11 viruses and the diagnosis of bronchiolitis in children admitted to a public hospital, as well as the presence of co-infection, the administration of antibiotics and oxygen therapy to those patients. Methods: A retrospective, descriptive study in which children aged between 0–13, treated at Hospital Municipal de Maringa in the State of Parana, Brazil, with acute respiratory symptoms, had nasopharyngeal swabs collected for virus research within the period of 1 year. Hospital records were analysed and the ones containing the diagnosis of bronchiolitis were selected. The following viruses were researched: Respiratory Syncytial Virus (RSV), Rhinovirus (RV), Adenovirus (AD), Enterovirus (EV), Parainfluenza types 1, 2 and 3, Metapneumovirus (MV),
Objective: To determine the CF severity for 3 scores [Bhalla, Kanga and Shwachman-Kulczycki]. Scores were performed by 2 medical professionals by double-blind technique. There was considered the mean between the evaluators and, in case of discrepancy, the data were evaluated by a third evaluator. Results: For distribution for the scores was: (i) Bhalla score: 8.81 ± 5.69; median = 8 (range 0 to 25); (ii) kanga score: 18.85 ± 5.86; median = 17 (range 10 to 40); (iii) Shwachman-Kulczycki: 66.24 ± 16.86; median = 65 (range 20 to 95). Regarding the association with genetic variants (modifier genes) of CF patients, we have: (i) Bhalla score was associated with 10 genes (11 polymorphisms per gene) on 125 genes were considered for association study. For genetic analysis, OpenArray™ platform (LifeTechnologies) was used. SPSSv22.0 performed the statistical analysis. alpha = 0.05. The achievement of clinical scores was performed considered published studied for each score [Bhalla, Kanga and Shwachman-Kulczycki] and maternal smoking during pregnancy were associated with disease severity, given that those cases had a mean hospital stay of 6.1 days, 2 admissions to ICU and oxygen therapy was needed in 62.5% of patients. Nevertheless, probably due to the reduced size of the sample, no statistical significance was found. Conclusion: RV and RSV were found to be the main etiologic agents for bronchiolitis in this population. In addition, it is suggested that RSV is related to greater severity.

097 BHALLA, KANGA AND SHWACHMAN-KULCZYCKI SCORE ON CYSTIC FIBROSIS: A LOOK BY THE MODIFIER GENES

Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertacchino (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Maria Angela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Adília Aparecida Dalbo Contrera Toro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Antônio Fernando Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); José Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas)

Objectives: To determine the CF severity for 3 scores [Bhalla (pulmonary disease by high-resolution computed tomography), Kanga (pulmonary exacerbation) and Shwachman-Kulczycki (assesses the overall clinical picture considering important aspects of the disease)] and to compare with the modifier genes variant with pulmonary expression associated with inflammatory response. Material: Included 126 CF patients with 2 CFTR mutations identified (classes I, II or III). Methodology: 256 polymorphisms per gene were assayed on 125 genes were considered for association study. For genetic analysis, OpenArray™ platform (LifeTechnologies) was used. SPSSv22.0 performed the statistical analysis. alpha = 0.05. The achievement of clinical scores was performed considered published published and maternal smoking during pregnancy were associated with disease severity, given that those cases had a mean hospital stay of 6.1 days, 2 admissions to ICU and oxygen therapy was needed in 62.5% of patients. Nevertheless, probably due to the reduced size of the sample, no statistical significance was found. Conclusion: RV and RSV were found to be the main etiologic agents for bronchiolitis in this population. In addition, it is suggested that RSV is related to greater severity.

098 EARLY PSEUDOMONAS AERUGINOSA COLONIZATION IN CYSTIC FIBROSIS: LONG TERM RESULTS OF AN ERADICATION PROTOCOL

Fabiola Villac Adde; Barbara Riqueira; Luiz Vicente R. F. Silva Filho; Cláudia M. A. Nakai; Marina B. Almeida; Joaquim C. Rodrigues; Pediatric Pulmonology Division, Instituto da Criança, Hospital das Clínicas – Medical School of USP

Antipseudomonal therapy at the time of early Pseudomonas aeruginosa (Pa) infection is used in cystic fibrosis (CF) patients in order to try to eradicate this pathogen but the optimal drug regimen is not established. Objective: Evaluate the rate of eradication of first Pa infection ever in CF patients that received systemic and inhaled antibiotics following a standard protocol. Successful eradication was defined as no Pa growth from airways samples in the 12 months following the eradication treatment (free of Pa). Patients and Methods: A retrospective review of the clinical records of CF patients followed at a pediatric CF reference center in their first Pa infection ever was performed. All patients were submitted to a standard eradication protocol between June 2004 and December 2012. The eradication protocol consisted of: step I (1st infection) = 3 weeks of ciprofloxacin or intravenous antibiotics + inhaled colistin 1,000,000 U bid for 6 weeks; step II (2nd infection) = 3 weeks of ciprofloxacin or intravenous antibiotics + inhaled colistin 1,000,000 U bid for 12 weeks; step III (3rd infection) = 6 weeks of ciprofloxacin + 6 months of inhaled colistin 1,000,000 U bid. Oropharyngeal swabs or sputum cultures were performed at least quarterly. Results: 29 patients, 11 female (38%), median age at first Pa isolation of 2.7 years (age range 3 m to 13.4 y) were evaluated. Four patients received only step I, 24 received step II and 13 received step III treatment. Twenty one (72%) patients were free of Pa at the end of the follow up: 4 after step 1, 10 after step 1 plus step 2 and 7 after step 1, plus steps 2 and 3. Conclusions: The first Pa infection occured in very young patients. This long term follow up demonstrated that most patients with early Pa infection may need more than one eradication treatment to become free of Pa infection. A more rigorous definition of successful Pa eradication was used in this retrospective analysis compared to other studies.

099 EVALUATION OF RISK FACTORS ASSOCIATED TO THE LEVEL CONTROL AND SEVERITY OF ASTHMA IN CHILDREN AND ADOLESCENTS

Natasha Yumi Matsunaga; Maria Angela Gonçalves de Oliveira Ribeiro; Emília da Silva Gonçalves; André Moreno Morcillo; José Dirceu Ribeiro; Adília Aparecida Dalbo Contrera Toro; UNICAMP

Objective: To assess risk factors associated with the level control and severity of asthma in children and adolescents. Methods: Children and adolescents with asthma were selected (7–17 years of age) from the Pediatric Pulmonology Outpatient Clinic of the State University of Campinas, Brazil. The attendees answered to the Asthma Control Test (ACT) to evaluate level of asthma control; and the questionnaire based on Global Initiative for Asthma (GINA) for severity classification and the International Physical Activity Questionnaire (IPAQ) to quantify physical activity. In order to categorize risk factors, we applied a structured questionnaire about origin, gender, personal and family history and environmental exposures. Chi-square, Fisher-Freeman-Halton tests were applied and odds-ratio were estimated (p < 0.05). Results: The mean age of the patients was 11.22 ± 2.91 years, with a median of 11.20 (7.00–17.60) years. Out of the 100 patients, 27, 33, and 40 were classified as having controlled asthma (CA), partially controlled asthma (PCA) and uncontrolled asthma (UA), respectively. As for asthma severity; 34, 19, and 47 were classified as having mild asthma (MA), moderate asthma (MoA) and severe asthma (SA), respectively. The chance of being in the UA Group was 3.74 times higher in patients with rhinitis. The presence of a smoker caregiver and maternal smoking during pregnancy were associated with disease severity.

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society. We found an association between exposure to dust and pets with the level of asthma control (p = 0.011 and p = 0.008) and severity (p = 0.021 and p = 0.007). There were no associations between the level of asthma control and severity with gender, origin and physical activities. Conclusions: Rhinitis was associated with worse asthma control and contact with cigarette smoke with disease severity. The contact with dust and pets were both considered risk factors for worse control and greater severity of asthma.

100 TRENDS IN THE HOSPITALIZATIONS FOR ACUTE BRONCHIOLITIS IN BRAZIL, 2008-2014

Kanama Tumba (FAMED, PUCRS); Talitha Camara (Instituto de Pesquisas Biomedicas, PUCRS); Leonardo Araujo Pinto (Instituto de Pesquisas Biomedicas, PUCRS)

Objective: to assess the temporal trends in the national incidence of admissions for acute bronchiolitis (AB) in Brazilian infants. We analyzed data from infants < 1 year of age before and after the introduction of the immunization with palivizumab. Material and Methods: an ecological study of a sample representative of the whole nation has been conducted. Data from infants hospitalized with acute bronchiolitis were analyzed. The analysis of the database of the Brazilian Public Health System (www. datasus.gov.br) was used to assess the absolute numbers of the infants hospitalized with bronchiolitis, using international classification of Diseases (CID 21), in the period between 2008 and 2014. Primary outcomes were the incidence of bronchiolitis admissions. We compared the temporal trends of hospitalization before and after implementation with palivizumab. Results: Between 2008 and 2014, there were 227,070 infants hospitalized with bronchiolitis. 60% of these infants were male. The incidence of bronchiolitis admissions increased over in the period 2008–2013 by 8.5 to 12.5 per 1000 person-year. However, from 2013 to 2014, the rate of incidence of AB decreased from 12.5 to 11.5 per 1000 person-year, after the implementation of immunization with palivizumab. Conclusion: there has been a progressive increase in the annual incidence rate of infants hospitalized with bronchiolitis between 2008 and 2013. By contrast, since 2014 after the introduction of palivizumab, there has been a reduction in the incidence rate of acute bronchiolitis in infants. Keywords: bronchiolitis, admission, incidence, trends, palivizumab.

101 RELATION BETWEEN RESPIRATORY ASSESSMENT PARAMETERS AND FUNCTIONAL CAPACITY OF CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS

Ana Carolina da Silva Almeida (Hospital Infantil Joana de Gusmão); Francieli Camila Mucha (Universidade do Estado de Santa Catarina); Renata Gonçalves Maba (Universidade do Estado de Santa Catarina); Gustavo David Ludwig (Universidade Federal de Santa Catarina); Elisana Barbosa (Hospital Infantil Joana de Gusmão); Camila Isabel dos Santos Schiwinski (Universidade do Estado de Santa Catarina)

Objective: To relate respiratory assessment parameters (spirometry and oscillometric) with the performance in the assessment test of functional capacity TGlitter-P in children and adolescents with cystic fibrosis (CF).

Materials and methods: cross-sectional observational analytical study with the participation of CF patients followed in Florianópolis / SC- Brazil. Clinical stability was insured by the application of two clinical scores (CPCs and 11 signals). Spirometry and impulse oscillometry were executed in Pneumotachograph Jaeger Master Scope IOS (Germany) according to the norms of the American Thoracic Society, they were obtained the following parameters: FEV1, FVC, FEV1/FVC, Z, R5, R20, X5, Fres e AX. Then participants performed the TGlitter –P, repeated after interval of 30 minutes, being considered the best performance test. TGlitter –P is in transit through a circuit that involves: up and down stairs, move objects on a shelf, walking and sitting and standing, carrying a backpack. Its performance is defined by step through the loop five times as fast as possible. Statistical analysis was conducted using SPSS 20.0 software. There was data normality the Shapiro-Wilk test and applied the Spearman correlation test with significance level of 5 % (p < 0.05). Results: participated 21 children / adolescents, 12 girls, and mean age of 9.59 ± 2.27anos (between 9 and 15 years). Patients showed spirometric values in percentage of predicted FVC: 85.52 ± 19.80% and FEV1: 68.47 ± 21.70%. There was no correlation between the time spent in TGlitter –P and percentage of predicted values of the spirometric variables and oscillometric (p > 0.05). However the absolute values of R5 (p = 0.039) and R 20 (p = 0.044) were correlated with the time spent in the test. Conclusion: children / adolescents with CF showed significant correlation between performance on TGlitter- P and absolute values of representative oscillometric parameters resistance (R5 and R20).

102 SEGREGATING FOR HUMANIZING

Ana Carolina Dias Galliati (Clinical nurse outpatient pediatric medical and surgical specialties of the Hospital of Ribeirão Preto); Simone de Mattos Garcia (Clinical nurse outpatient pediatric medical and surgical specialties of the Hospital of Ribeirão Preto); João Luiz Manfredini Neto (Clinical nurse outpatient pediatric medical and surgical specialties of the Hospital of Ribeirão Preto); Albin Eugenio Augustin (The medical assistant pediatric pulmonology department of the University Hospital of Ribeirão Preto); Ieda Regina Del Ciampo (The medical assistant pediatric gastroenterology department of the University Hospital of Ribeirão Preto)

Introduction: Cystic fibrosis is a lethal inherited disease. Bacterial infection of acquiring bacteria contributes significantly to prognosis. Objective: The study proposes a hospital circulation strategy to reduce cross-transmission in cystic fibrosis. Methodology: HC Criança has 4 waiting rooms where patients remain waiting for health care routine. Despite colonized and non-colonized patients are scheduled in different days, there are situations when patients ended up together, because of exacerbations or rescheduled appointments. Given the importance of this, it was suggested by medical and nursing team, that this segregation would happen in a humane way. The applied proposal was: initially the nursing and the medical team, analyze the days listing of patients and put on a database and differentiate who is colonized and the kind of bacteria. Upon arriving at the clinic, the patient passes for reception and is forwarded to biometry room, where the nurses already have the listing with this separation. In this place, the records receive color markings according to the infecting bacteria. After the biometry, the patient is directed to different waiting rooms, according to the kind of colonization. The doors of treatment rooms receive the same marks found on the medical records. Thus, the multidisciplinary team has guidelines to call patients only in offices with the same marks seen in the records. After finishing, the room is cleaned with alcohol 70%. Results and Conclusions: There was excellent acceptance of patients for this segregation and was a former request of them. Doctors acceptance was also good, without difficulties to change to a new room to examine other patient. The only problem was with some non-medical professionals, because they were very used to have their own offices. However, gradually acceptance also occurred. In the long run, we will analyze the cross-infection rates, comparing them to previous data obtained in the same reference hospital.

103 EFFECT OF THE USE OF VIDEOS DURING REHABILITATION PROGRAMS ON PERCEIVED EXERTION IN CHILDREN WITH CHRONIC PULMONARY DISEASE

Natália Galap Sorges; Daniel Zeniteno Araos; Hospital Guillermo Grant Benavente

Objectives: The aim of this study was to investigate the effect of using videos during aerobic training on perceived exertion in children with...
chronic pulmonary disease. Materials and patients: 15 patients were included, ages: 10 (5–17), diagnostic: Asthma (4), Bronchiolitis obliterans (5), Duchenne muscular dystrophy (3), Cystic Fibrosis (2) and Severe Scoliosis (1). Reflective cones, FDA approved pulse oximeter, EPINfant printed scale, treadmill and a tablet as video player. Methodology: Patients performed the 6 min walk test (6MWT). One week later, they performed a constant load test in treadmill with 60% of the average speed in the 6MWT. After two weeks, they performed the same test but using videos of their preference during the test. Dyspnoea and Legs Fatigue were analized using the 0–10 scale EPINfant, using and not using videos. For the statistical analysis, we used the non parametric Wilcoxon Test with a 95% confidence interval. Results: There was a significant difference (p < 0.05) in the averages of Dyspnoea (from 4.9 to 2.7) and Legs fatigue (from 4.7 a 2.8). Conclusions: A significant difference can be observed in perceived exertion in children under 18 years old, with chronic pulmonary disease when they use videos during aerobic training. This could be a useful resource to improve physical performance in children during rehabilitation protocols.

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104 ATYPICAL PNEUMONIA IN CHILDREN DIAGNOSED WITH IMERSLUND GRÄSBECK SYNDROME

Moeda Lunelli; Adriano Nori Rodrigues Taniguchi; Debora Quiorato Fernandes; Gilberto Bueno Fischer; Helena Terezinha Mocelin; Hospital da Criança Santo Antonio – Santa Casa de Misericórdia de Porto Alegre

Objective: To report a case of atypical pneumonia in patient with pancytopenia and hemolytic anemia, alerting doctors to this rare condition.

Case Report: Five year-old girl, hospitalized for cough, fever and decreased urine output for a week. She was pale, emaciated, with tachypnea and respiratory effort. CBC: hemoglobin 7.6 g/dL, MCV 106fL, leucocytes 1250/mm³ (eosinophils 1.6%; basophils 1.5%, lymphocytes 68.8%, neutrophils 21.4%, monocytes 6.7%), platelets 41,000/mm³, reticulocytes 13%. Urine test with proteinuria. Chest X-ray with consolidation in the left upper lobe. Cefuroxime was started; cotrimoxazole was added due to the risk of an opportunistic infection (persistent pancytopenia). Revising medical records, there was a previous diagnosis of vitamin B12 deficiency. Imerslund Gräbeck Syndrome was suspected. New urine test confirmed proteinuria, and CBC showed agglutination of red blood cells. Direct Coombs was positive for C3 suggesting the cold antibody; MCV was 14 fL and the immature platelet fraction was 42% suggesting increased bone marrow regeneration (characteristic of autoimmune pancytopenia). Mycoplasma IgG and IgM were positive. Macrolide was started and treatment with B12 vitamin was added with good therapeutic response. DISCUSSION: Many times infection by Mycoplasma produces autoantibodies against hematopoietic cells, however rarely develops to autoimmune pancytopenia and leukemoid reaction. In this patient, consumption by infection and impaired hematopoiesis due to B12 vitamin deficiency may have contributed to the low white blood cell count and severe neutropenia. The Imerslund Gräbeck is a rare syndrome characterized by megaloblastic anemia and asymptomatic proteinuria. In severe cases of B12 vitamin deficiency, pancytopenia is observed by failure of cell division where it works as a cofactor. The recommended treatment is B12 vitamin replacement therapy.

105 CLINICAL AND EPIDEMIOLOGICAL CHARACTERIZATION OF CYSTIC FIBROSIS IN A PEDiatric CENTER OF EXCELLENCE IN NORTHEASTERN BRAZIL

Milena Silva Reis (School of Medicine of Bahia, Federal University of Bahia); Paloma Herson Bittencourt (Federal University of Bahia); Bianca Silva Sampeio (Federal University of Bahia); Luis Ribeiro Mota (Federal University of Bahia); Renata Lúcia Leite Ferreira Lima (Federal University of Bahia); Edna Lúcia Santos de Souza (Federal University of Bahia)

Objectives: To describe the clinical and epidemiological characteristics of the cystic fibrosis patients receiving care at the pediatric multidisciplinary centre. Material and Methods: This was a cross-sectional study. Cystic fibrosis (CF) patients under the age of 21 years followed up at outpatient pediatric clinics between March 2005 and August 2015 were included. The data were obtained from medical records followed by blood sample collection from the patients for molecular analysis for Phe508del, G542X, G551D, R553X, R1162X and 3120+1G>A mutations by standard methods. There was a descriptive analysis of the data registered. Results: Fifty-four patients were included, 51.9% were male. All the children were non-white. The median age of the patients at admission to the study was 10.0 years. The median age at onset of symptoms and at disease diagnosis was 3.5 months and 4.8 years, respectively. At diagnosis the main clinical manifestations recorded were: respiratory symptoms (79.6%), difficulty in gaining weight (77.8%) and steatorrhoea (51.9). None of the patients had meconium ileus and hepatobiliary disease occurred in five cases (9.3%). Four related patients were excluded of genetic analysis. The Phe508del mutation was detected in 36% (18/50) of the patients, allele frequency of 27% (27/100), G542X was found in three cases, 3120+1G-A was presented by three and R1162X was identified in one patient. The mutations G551D or R553X were not observed. Fourteen participants (28%) already have the two alleles mutations detected, six (12%) remain with a genotype partially known. Conclusions; The disease was diagnosed at a late stage. The majority of the cases presented classical symptoms of disease. The meconium ileus was not observed and there was a high frequency of hepatic disease. The frequency of the Phe508del mutation was lower and the 3120+1G-A, an African mutation, was observed, which reflects the high degree of miscegenation in the study population.

106 CASE REPORT: INHALED MERCURY POISONING IN A PEDIATRIC PATIENT

Linda Andrea Betancur (Hospital San Vicente Fundación); Silvia Palacio Petri (Hospital San Vicente Fundación); Olga Lucía Morales Muñera (Universidad de Antioquia y Hospital San Vicente Fundacion); María Gabriela García (Universidad de Antioquia); Andrea Catalina Cardona Moreno (Universidad de Antioquia)

Objective: To report a case of inhaled mercury poisoning in a child. Methodology: case report. Results: 15-month-old girl, with no relevant past; her stepfather works in a gold mine. The stepfather, the mother and the child began with respiratory symptoms after burning a piece of gold within the house. A day later, the stepfather appeared with mild symptoms. The mother was admitted to the hospital with moderate symptoms, and the high resolution chest tomography (HRCT) showed diffuse ground-glass opacities of the upper lobes. The girl required admission with fever, diarrhea and marked respiratory distress. The HRCT showed right pneumothorax and bilateral generalized consolidation, she evolved to an acute respiratory distress syndrome (ARDS), requiring high ventilatory support. Mercury blood and urine levels were identified: 1050 mcg/L (normal value 20 mcg/L) and 300.99 (normal value 50 mcg/L), respectively. Penicillamine, N-acetylcysteine, EDTA and garlic were used as chelating agents. In addition, methylprednisolone boluses were used. The child had a slow evolution toward improvement and was extubated to a nasal cannula. Conclusions: In countries where illegal mining is as frequent as in Colombia, inhaled mercury poisoning should consider in patients presenting to the emergency department with sudden respiratory impairment of varying severity.

107 A NEGATIVE REPORT TO MODIFIER GENES OF THE CYSTIC FIBROSIS: THE CASE OF –173G>A POLYMORPHISM IN THE MIF GENE

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SPIROMETRIC CHANGES IN CHILDREN AND ADOLESCENTS WITH SICKLE CELL DISEASE

Tatiane da Annunciace Ferreira (Serviço de Pneumologia Pediátrica – Hospital Universitário Professor Edgard Santos-Uniuversidade Federal da Bahia); Região Tere-Ramos (Curso de Pós graduação de Medicina e Saúde Humana – Fundação Bahiana para Desenvolvimento das Ciências); Luisa Danielle Alves de Souza Santos (Academia da Faculdade de Medicina da Universidade Federal da Bahia); Salvador Henrique de Souza Miranda (Academia da Faculdade de Medicina da Universidade Federal da Bahia); Renata Maria Pereira Vieira Barbosa (Academia da Faculdade de Medicina da Universidade Federal da Bahia); Ana Marice Teixeira Ludue (Doutora em Medicina e Saúde Humana, Professora Adjunta da Escola Bahiana de Medicina e Saúde Pública

Objective: To describe spirometric indices and lung function patterns of children and adolescents with Sickle Cell Disease (SCD). Material: this study involved 58 HbSS participants, aged between 6 and 10 years old, and 42 healthy patients as comparison group. Spirometry was performed according the American Thoracic Society and the Brazilian Thoracic Society and pulmonary impairment was classified as mild (FEV1 60–79% predicted), moderate (FEV1 41–59% predicted) or severe (FEV1 < 40% predicted) Methods: cross-sectional study; patients were recruited from three SCD outpatient clinics. Demographic and clinical data were recorded. Data analysis was performed in SPSS version 21.0; Pearson correlation were done and considered P-value < 0.05 as the significance. Results: children with SCD were 62,1% males, 94,8% nonwhites and aged 12,47 ± 3,12 years while controls were 33,3% males, 85,4% nonwhites and aged 11,33 ± 3,19 years. Body Mass Index were normal in 87,2% of cases and 61,5% of controls, while 12,9% of cases had slimness and 26,9% of controls were overweighted. All spirometric indices had mean within normal range, but children with SCD had smaller parameters compared with controls. Cases and controls, respectively: Forced expiratory volumes (FEV1) pre 81,22 ± 16,19 vs. 98,81 ± 13,41 (p < 0.0001); Forced Vitalcapacity (FVC) pre 79,71 ± 16,68 vs. 95,29 ± 14,19 (p < 0.0001); FEV1/FVC pre 102,48 ± 4,91 vs. 104,50 ± 4,37 (p < 0,036). When we separated cases into children between 6 and 9 years and 10 and 18 years, the older had smaller indices (cases 6 a 9 anos e cases 10 a 18 anos, respectively: FEV1/FVC pre 105,17 ± 2,58 vs. 101,78 ± 5,14; p < 0,003); (FEF 25-75% 114,42 ± 37,18 vs. 94,98 ± 22,07; p < 0.023). Results of spirometry were classified as obstruction (controls and cases, respectively: 8,6% x 0%), restriction (37,9% x 9,5%), and normal lung function (43,1% x 85,7%). Conclusion: The majority of cases has abnormal lung function and children and adolescents with SCD have smaller indices spirometrics compared to comparison group.

108 OXYGEN SATURATION, PERIODIC BREATHING AND APNEA DURING SLEEP IN INFANTS 1 TO 4-MONTH OLD LIVING AT 2,560 METERS ABOVE SEA LEVEL

Santiago Uccro (Fundación Sante Fe de Bogota – Colombia); Claudia Granados (Universidad Javeriana – Bogota, Colombia); Karen Parejo (Clínica Shaio – Bogota – Colombia); Fernando Guillen (Hospital del Rio. Universidad del Azuay. Cuenca – Ecuador); Fausto Ortega (Hospital Luis Fernando Martinez. Cuenca – Ecuador); Sonia Restrepo (Hospital de La Misericordia – Bogota – Colombia)

Objective: To describe oxygen saturation (SpO2) levels during sleep in infants aged between 1 and 4 months living at an altitude of 2.560 m. The secondary objectives were the description of periodic breathing (PB) and apnea indexes. Methodology: polysomnography was performed in 35 healthy infants 1–4 months in Cuenca (Ecuador) at 2.560 m. Results: the median for SpO2 was 92% and 4.9% for PB. The median for the central apnea index was 23.7/hour and 15.4/hour when related to PB. No correlation was found between PB and SpO2. Conclusions: SpO2 was lower than the values at sea level and PB and central apnea index were higher. When apneas associated with PB were not considered, the central apnea index was similar to that found at sea level. The absence of correlation between PB and SpO2 suggests that low SpO2 is attributable to the decreased inspired PO2 characteristic of high altitudes and not to the increase in PB. At this altitude the normality threshold for the SpO2 in young infants should be differentiated according to wakefulness or sleep stages. This does not occur at sea level or low altitude, where a single value is sufficient for both of these circumstances.

Pediatric Pulmonology

109 RISK FACTORS ASSOCIATED WITH ASTHMA DIAGNOSIS IN A COHORT OF CHILDREN WITH RECURRENT WHEEZING

Vitória Marino Dobbario De Paula (Pontificia Universidad Católica De Campinas); Adélia Aparecida Dalbo Contraer Toro (UNICAMP); José Dirceu Ribeiro (UNICAMP); Natasha Yumi Matsuosnaga (UNICAMP)

Objectives: To analyze risk factors associated to asthma diagnosis in a cohort of children that had recurrent wheezing among the first two years of live. Methods: This study investigated medical files of children from the outpatient clinic of wheezing infants of the HC UNICAMP that were born between the years of 2006 and 2009. Data were collected through a questionnaire with information about personal and family background, wheezing medical history and complementary exams. Family history of atopy associated to changes on IgE levels or eosinophils were used to establish atopy diagnosis. The statistical tests used were chi square, Fisher, Mann-Whitney. Data was stored on SPSS 16 and was used significant p below 0.05. Results: 180 medical reports with complete information about atopy were analyzed and 60% were male patients. 144 were considered atopic while 66 as non-atopic. Data analysis indicated that atopic patients
Methods: 90 patients (Median age = specific P.a IgG antibodies to early detect the P.a chronic lung infection.

Objectives: To evaluate the capacity of a serological test for detection of Emilio Levy; CACYF Group; Universidade Estadual de Campinas

Cystic Fibrosis by a Serological Test

ORAL MONTELUKAST VERSUS INHALED BECLOMETHASONE IN THE PROPHYLACTIC TREATMENT OF POST ACUTE VIRAL BRONCHIOLITIS WHEEZING

Carolina Andrade Neves Silva; Luciana Zignago Moreira dos Santos; Chalene Guimarães Soares Mezzicôncio; Wilson Rocha Filho; Hospital Infantil João Paulo II

Objective: We aim to determine the potential benefit of inhaled beclomethasone dipropionate (BDP) and oral montelukast (MK) in the natural history of post acute viral bronchiolitis (AVB). Materials and Methods: This is an open, prospective, randomized, parallel control study, involving infants of 1 year of age or younger hospitalized for AVB. All patients with moderate-severe bronchiolitis (Wang score equal or superior to 8) were eligible for the study. Sixty seven infants diagnosed with AVB between July 2013 and May 2015 were selected for the study. Thirty children were excluded either because they fulfilled exclusion criteria or lost their follow up. At hospital discharge, recruited patients were randomly assigned into 3 study groups: no drug, MK or BDP and schedule for monthly follow-up appointments for 6 months. The primary variables were: need for readmission and number of visits to the emergency department (ED). Secondary variables were: number of asymptomatic days and need of bronchodilator and/or oral corticosteroid. Results: The analysis was performed on 37 children, 12 randomized to receive no prophylaxis, 18 randomized to MK and 7 to use BDP. There were 3 hospitalizations in children with no prophylaxis and 3 receiving MK. There were no readmissions in the group receiving BDP. Visits to ED occurred in 6 children with no prophylaxis, 15 in those receiving MK and in 6 children receiving BDP. There was no statistics difference between the groups considering readmission, visit to the ED, asymptomatic days after discharge, bronchodilator and oral corticosteroids use. Conclusions: Our study showed no benefit of MK or BDP in the treatment of post AVB. A larger number of patients will allow us to establish more reliable statistical data.

112 EARLY DIAGNOSIS OF PSEUDOMONAS AERUGINOSA CHRONIC PULMONARY INFECTION IN PATIENTS WITH CYSTIC FIBROSIS BY A SEROLOGICAL TEST

Renan Marrichi Mauch; Claudio Lucio Rossi; Talita Bianchi Aiello; Carlos Emilio Levy; CACYF Group; Universidade Estadual de Campinas

Objectives: To evaluate the capacity of a serological test for detection of specific Pa IgG antibodies to early detect the Pa chronic lung infection. Methods: 90 patients (Median age = 8,93 years) without Pa chronic infection (according to the Leeds criteria) were followed. Serum samples were collected from these patients in different periods of time and the IgG dosage was made by a previously standardized ELISA technique, with the polivalent antigen St-Ag:1–17. A cut-off value of 15.30 U/mL determined a positive result for the serological test. Results: 17 patients had positive IgG levels in the baseline; 09 patients became chronically infected and all of them had positive anti-Pseudomonas IgG levels in the baseline (Median = 17.05 U/mL), which were significantly higher than those of patients who didn’t develop chronic infection (Median = 4.33 U/mL) and remained significantly higher until the end of the follow-up (152.30 vs. 10.63 U/mL). A positive result for IgG in the baseline was a risk factor for the development of Pa chronic lung infection (OR = 4.91; p < 0.05). Only 03 patients acquired Pa in culture without positive IgG levels, and 27 patients did not acquire Pa in culture but had positive IgG levels during the study. Conclusions: Our results support the literature, which shows that the transition from Pa intermittent colonization to chronic infection is followed by an enhanced specific IgG response. The 27 patients who enhanced the IgG levels without changing the microbiological status are probably chronically infected by Pa, but this infection must have not been detected by microbiological culture. Although the results are only preliminary, the inclusion of anti-Pa IgG serology in the CF diagnostic routine, as a complementary test to the microbiological respiratory culture, must be considered, so Pa chronic lung infection can be early diagnosed.

113 PLEURAL TUBERCULOSIS IN CHILDREN AND ADOLESCENTS: WHEN SHOULD WE THINK OF THIS DIAGNOSIS?

Maílva Lunelli (Universidade Federal do Rio Grande do Sul – Programa de Pós Graduação em Ciências Pneumológicas); Cristiano Feijó Andrade (Hospital da Criança Santo Antônio – Santa Casa de Misericórdia de Porto Alegre); Helena Teresinha Moczelin (Hospital da Criança Santo Antônio – Santa Casa de Misericórdia de Porto Alegre); João Antônio Bonfadini Lima (Hospital da Criança Santo Antônio – Santa Casa de Misericórdia de Porto Alegre); Rodrigo Moreira Bello (Hospital da Criança Santo Antônio – Santa Casa de Misericórdia de Porto Alegre); Gilberto Bueno Fischer (Hospital da Criança Santo Antônio – Santa Casa de Misericórdia de Porto Alegre e Universidade Federal de Ciência da Saúde de Porto Alegre)

Objective: Describe the key signs and symptoms observed in patients with pleural tuberculosis (TP) diagnosed in our department. Material: We describe the findings of patients undergoing PT, treated at a hospital in southern Brazil between 2007 and 2014. Methodology: This is a descriptive, case series study of children and adolescents with PT diagnosis confirmed through biopsy showing caseous granuloma. Results: Between 2007 and 2014 there were 39 patients admitted to the service aged 1 to 17 years (mean: 11.1; median: 12 years). The most frequent complaints were: cough (67%), fever (67%), chest pain (59%), weight loss (31%), dyspnea (15%) and night sweats (13%). Chest radiography showed right pleural effusion in 64% of cases. The median time between onset of symptoms and diagnosis was 16 days (median: 11 dias). Before admission 74% used antibiotics for pneumonia. During hospitalization, information was obtained about the form of contagion, which 66% of patients reported having prior contact with tuberculosis patients. Conclusions: There was a delay in diagnosis and inappropriate use of antibiotic in most patients. Predominant symptoms were cough, fever and thoracic pain (indistinguishable from pneumonia findings). In non-toxemic patients with pneumonia with therapeutic failure, tuberculosis contact should be investigated. Early diagnosis of pleural tuberculosis is challenging, and its delay may cause unnecessary use of antibiotics.

114 ASPECTS OF SURGICAL TREATMENT OF ASPHYXIATING THORACIC DYSTROPHY

Débora Quiorato Fernandes; Débora Quiorato Fernandes; Gilberto Bueno Fischer; Julio de Oliveira Espinel; Angelica Barba Rueda;

Pediatric Pulmonology
Objective: To describe the experience and the challenges for treating an abnormality in the development of the chest wall in a Paediatric Thoracic Surgery department. Methods: 9 medical records from patients with asphyxiating thoracic dystrophy at any age. Results: 3 patients died due to septicemia in the immediate postoperative period. Conclusion: Treatment of patients with asphyxiating thoracic dystrophy is complex due to the conditions associated with the patients. Even with optimal treatment, results are not very encouraging. The lateral thoracic expansion is a feasible procedure and should be considered for the treatment of asphyxiating thoracic dystrophy at any age.

115 INCIDENCE AND TREATMENT OF METHICILLIN-RESISTANT STAPHYLOCOCCUS AUREUS INFECTION IN CYSTIC FIBROSIS PATIENTS: A COHORT STUDY

Paloma Horeis Bittencourt (School of Medicine of Bahia, Federal University of Bahia); Carlos Sidney Silva Pimentel (School of Medicine of Bahia, Federal University of Bahia); Bianca Sampato Bonfim (School of Medicine of Bahia, Federal University of Bahia); Carolina de Godoy Abmeida (Universitary Hospital Professor Edgard Santos, Federal University of Bahia); Paulo José Cauduro Marostica (Federal University of Rio Grande do Sul); Edna Lúcia Santos de Souza (School of Medicine of Bahia, Federal University of Bahia)

Objectives: To determine the incidence of the respiratory tract colonization by methicillin-resistant Staphylococcus aureus (MRSA) and investigate the identification of the bacterium after the use of routine treatment in patients with Cystic Fibrosis (CF). Material and Methods: It was a retrospective cohort study. CF patients under the age of 21 years with CF confirmed by sweat test or genetic study followed up at outpatient clinics in a Teaching Hospital were included. The data were obtained from medical records. The sputum samples or oropharyngeal swabs were collected from patients for microbiological culture. The colonization or infection in the respiratory tract of patients occurred from January 2008 to June 2015 were registered. After 2013, all the subjects with MRSA isolated were treated with the standard treatment and side effects were not observed. Only one child had new isolation of MRSA during follow-up (after 20 months). Conclusions: The incidence of MRSA infection was high and it occurred in young patients. The therapeutic regimen was effective and safe and it can be a good option to treat MRSA infection.

116 CASE REPORT: SINGULAR EVOLUTION OF CONGENITAL PULMONARY LYMPHANGIECTASIA
Renata Wrobel Flesca Coelho; Déborah Aragão de Pinho Silveira; Tania Wrobel Flesca; Renato Farn D’Amoed; Patrícia Fernandes Barreto Machado Costa; Laurinda Voko Shinzato Higa; Instituto Nacional de Saúde da Mulher, da Criança e do Adolescente Fernandes Figueira (IFF/ FIOCRUZ)

Objective: Describe diagnostic approach and peculiar evolution of a patient with congenital pulmonary lymphangiecitsia (CPL). Case Report: Male full term infant presented early respiratory distress requiring respiratory support and oxygen therapy, Chest radiograph showed pulmonary cysts and CT scan done at 13days revealed diffuse parenchymal involvement, mainly lower lobes and diffuse interstitial thickening. Patient maintained tachydyspnea and PCO2 50-70 mmHg, requiring continuous oxygen therapy and intermittent non-invasive ventilation. Respiratory function tests revealed diminished complacence and elevated resistance, with bronchodilator response. Cystic fibrosis, alpha 1-antitrypsin deficiency, infectious, renal, neurologic and congenital heart diseases were ruled out. Open-lung biopsy was performed on the 58th day of life and the findings were consistent with congenital pulmonary lymphangiectasia. Hospital discharge occurred at 5mo with continuous oxygen therapy and nocturnal non-invasive ventilation. At 3yo, oxygen therapy was discontinued and multidisciplinary follow up was maintained. Currently at 16yo, the patient presents body mass index below 3rd percentile, effort dyspnea and SatO2 93–95% at rest. He uses regularly long acting ?-agonist and inhaled corticosteroids, maintains CO2 retention (50-68 mmHg) without hypoxemia. 2013 CT scan showed hyperinflation and interlobular septal thickening, without pleural effusion. Regarding pulmonary function tests, he maintained the same pattern, since 2009, with obstructive pulmonary disease, with no decline on functional parameters. Conclusion: Symptomatic CPL since newborn usually presents high mortality rates. Dilated lymphatics and chylothorax lead to hypoplastic lungs and respiratory failure. The present study describes a 16yo patient with neonatal respiratory symptoms and histopathological findings suggesting CPL. This case highlights the importance of thinking about CPL diagnosis and that survival is possible.

117 ASSOCIATION BETWEEN HYPOVITAMINOSIS D AND FREQUENCY OF PULMONARY EXACERBATIONS IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS
Renata Orsoguatta (Centro INFANT, Instituto de Pesquisas Biomédicas PUCRS); Paulo Marostica (UFRGS); Leonardo A. Pinto (PUCRS)

Objective: In this study, we evaluated the association between vitamin D levels and nutritional status, pulmonary function (PF) and pulmonary exacerbations (PE) in children and adolescents with cystic fibrosis (CF). Material and methods: 25-hydroxyvitamin D (25(OH)D) levels of 37 children and adolescents were evaluated retrospectively. Pulmonary function data, albumin, body mass index (BMI), height for age (H/A) and frequency of pulmonary exacerbations were assessed. Vitamin D levels were measured and divided into two groups: normal (≥ 30 ng/ml) and insufficient/deficient (< 30 ng/ml). Results: hypovitaminosis D (25(OH)D <30 ng/ml) was observed in 54% of patients. The mean of 25(OH)D was 30,53 ± 12,14 ng/ml. Pulmonary function and nutritional status were not associated with vitamin D levels. The number of pulmonary exacerbations over a period of two years (p = 0.007) and post-dosing period of 25(OH)D (p = 0.002) was significantly higher in patients with hypovitaminosis D. There was a trend of lower 25(OH)D levels during autumn and winter.
ACHROMOBACTER XYLOSEXIDANS PULMONARY INFECTION IN PATIENTS WITH CYSTIC FIBROSIS: EPIDEMIOLOGY AND ANTIMICROBIAL RESISTANCE IN 12 YEARS OF FOLLOW-UP IN A REFERENCE CENTER

Renan Marrichi Mauch; Tatiana Bianchi Aiello; Antonio Fernando Ribeiro; José Dirceu Ribeiro; Carlos Emilio Levy Universidade Estadual de Campinas

Objectives: To evaluate the prevalence of positive microbiological respiratory cultures to A. xylosodans and their patterns of antimicrobial resistance in 12 years of follow-up (2002–2014) of CF patients in the reference center of the HC-Unicamp. Methods: In each year, a minimum of 04 respiratory samples was collected from each patient followed in the HC-Unicamp CF center and the sensitivity tests were performed according to the CLSI (Clinical & Laboratory Standards Institute – Wayne, PA, United States) recommendations. Results: A total of 56 patients presented 192 positive samples during the period, being 29 (52%) of female gender. There was an enhancement in the number of cases from 2005 to 2008 (3 to 15 patients), followed by a sharp drop from 2009 to 2011, which was succeeded by a new enhancement from 2012 to 2014 (4 to 12 patients). This last period concentrated 45% of the 192 positive samples, with continuous enhancement in their number (24 to 36 samples) and substantial enhancement in the number of chronically infected patients (n = 10). In 12 years, it was observed a considerable percentage of resistance to amikacin (38%), cefepime (33%), ciprofloxacin (31%) e sulphamethoxazole-trimethoprim (22%). Conclusions: The prevalence of A. xylosodans is low, considering that our Center attends about 250 patients, and was quite variable over time. However, it is worthy the enhancement observed in the last 3 years, which is due to the increase in the number of chronic infection cases. Another concern is the high rate of resistance to the antibiotics previously cited, which reflects their large use in the treatment of the disease. In chronic infection, the rate of multiresistance is higher, favouring the bacterial adaptation in the airways and hampering the infection treatment, which leads to a rapid deterioration in the lung function. Thus, the frequent monitoring of infection is essential for its early detection and better clinical management.

EVALUATION OF GROWTH MEASUREMENTS IN THE FIRST TWO YEARS OF LIFE IN CHILDREN DETECTED WITH CYSTIC FIBROSIS NEONATAL SCREENING

Denise Tieni Miyakawa (Hospital Infantil Pequeno Príncipe); Jocemara Garmini (Hospital Infantil Pequeno Príncipe); Mirella Aparecida Neves Almeida (Hospital Infantil Pequeno Príncipe); Dyrlanne Marcia Lopes Bastos (Hospital Infantil Pequeno Príncipe); Gleison Luiz Picharski (Instituto De Pesquisa Peté Pequeno Príncipe)

Patients with Cystic Fibrosis (CF) are a challenge for every Multidisciplinary Team and the nutritional support is an import point in their treatment. CF children should have normal growth and they require appropriate energy and nutrients intake. According to the American Consensus report on nutrition for pediatric patients with CF (2002), there are three specific times when special attention should be focused: the first 12 months after the diagnosis, from birth to 12 months of life and peripubertal period. Growth and nutritional status should be monitored every 3 months and, during the first year, every month. Evaluate the growth measurements of CF children detected with neonatal screening from zero to 24 months. We reviewed the

records of 33 patients with cystic fibrosis, from 0 to 24 months of life in 6 moments (0, 3, 6, 12, 18, 24 months). Informations about age, length, weight, anthropometric classification (WHO), breast or formula feeding and complementary food was collected. Thirty three patients, 13 (39,3%) male, 20 (female); 26 (78,7%) with pancreatic insufficiency (PI) and 86,6% breastfed in the first evaluation (T0). In T0 (0–2 months) the patients with infant formula was in the worst nutritional classification (z-score W/A, L/A e BMI/A < -3). After the pancreatic enzyme replacement therapy was initiated and the adequacy of nutrition was started, the weight and length gain was similar between breastfed and formula fed group. At two years of life, all anthropometric classifications are out of risk of undernutrition ( z-score W/A, L/A e BMI/A > -2). The infants weight increased with a caloric intake higher than 150 Kcal/Kg/day and protein twice the recommended for age. The early nutritional intervention with adequate calories, nutrients and the replacement of pancreatic enzymes and vitamins are important for a positive weight gain and growth in CF infants and toddlers detected with CF newborn screening.

TRICHSOPHORON INFECTION IN A PATIENT WITH CYSTIC FIBROSIS – DOES IT WORSEN THE CLINICAL STATUS?

Jessica Ventura (Unidade de Pneumologia, Instituto da Criança HCFMUSP); Marcelo Bernasconi Daniel (Unidade de Pneumologia, Instituto da Criança HCFMUSP); João Nobrega de Almeida Junior (Divisão de Microbiologia, Laboratório Central HCFMUSP); Luiz Vicente Ribeiro F da Silva Filho (Unidade de Pneumologia, Instituto da Criança HCFMUSP)

Objective: To describe clinical and functional evolution following respiratory infection by Trichosporon sp. in a cystic fibrosis (CF) patient. Case Report: HCRM, male, 16 years old. Phe508del/G542X mutations, attending our outpatient CF Clinic since 2003. At the diagnosis (4 years old) he was already presenting symptoms of pancreatic insufficiency and lower respiratory tract symptoms, being treated accordingly: he started Pulmozyme at 5 years old and received several courses of eradication therapy for P. aeruginosa in the subsequent years. He became also colonized intermittently with B. cepacia complex. In 2014 he developed diabetes related to CF and insulin therapy was started. He is currently chronically colonized by Methicillin-susceptible S. aureus and intermittently colonized by P. aeruginosa. He used suppressive treatment with alternate-months of inhaled tobramycin from 2007 to 2014. Trichosporon sp. was first detected in a sputum culture in September 2014, when his FEV1 was 1.47L (46%) and his BMI 19.2 kg/m2. Sequential sputum cultures remained positive for Trichosporon sp. after that, which was phenotypically identified as a T. asahii. Further DNA sequencing of intergenic ribosomal DNA region (IGS-1) revealed 99% similarity with T. mycotoxivorans. During a routine clinical visit in July 2015 we decided to treat him with fluconazole for 21 days and sulfamethoxazole-trimethoprim for 14 days; although he reported to be well, his FEV1 dropped to 1.24L (37%). After treatment Trichosporon did not grow in subsequent sputum cultures, but his FEV1 remained stable (1.27L – 39%). Discussion: Trichosporon sp. is a novel pathogen in patients with CF and its pathogenic role is still unclear. T. mycotoxivorans seems to be the most prevalent species in CF patients, and fluconazole and voriconazole are the main therapeutic options. While this patient had a decrease in FEV1 after infection, we didn’t observe an improvement after specific treatment.

CORRELATION BETWEEN NASAL PATENCY VARIABLES AND CONE BEAM TOMOGRAPHY IN MOUTH BREATHERS

Raguel Harumi Uezumi Satto Sakai (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Sciences, State University of Campinas); Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Sciences, State University of Campinas)
Objective: To correlate the computerized tomography variable cone beam (CTCB), acoustic rhinometry (AR) and computed rhinonometry (CR) in mouth breathers (MB) with maxillary atresia. Material: Thirty mouth breathing children with ages ranging from 7 to 13 years old and maxillary atresia with posterior crossbite were studied. Methodology: A cross-sectional study was performed. The children underwent AR (volume 0-5 cm, volume 2-5 cm, minimum cross-sectional areas MCA1 and MCA2), CR [inspiratory resistance (I), expiratory resistance (E) and nasal air flow (F) on the left nostril (LN), right nostril (RN) and on the medium of both nostrils] and CTCB [pinform opening (width 1), head of the inferior turbinate (widths 2 and 3), head of the middle turbinate (widths 4 and 5) and maxillary width (width 6)] before (BVC) and after (AVC) use of vasoconstrictor. The data was compared by linear regression (alpha = 0.05).

Results: There was correlation of AR versus CTCB-BVC in the LN of MCA1 with width 3 (p = 0.009; R² = 0.222), CR correlated with CTCB-BVC in the LN of F with width 5 (p = 0.046; R² = 0.134) and the medium F of both sides with width 5 (p = 0.05; R² = 0.13). There was correlation of AR in LN-BVC: volume 0-5 cm with F (p = 0.019; R² = 0.182); volume 0-5 cm with I (p = 0.026; R² = 0.166); volume 2-5 cm with F (p = 0.019; R² = 0.182); volume 2-5 cm with I (p = 0.032; R² = 0.154); MCA1 with F (p = 0.026; R² = 0.164); MCA1 with I (p = 0.022; R² = 0.173); MCA2 with F (p = 0.021; R² = 0.177); MCA2 with I (p = 0.042; R² = 0.139) and LN-AVC: volume 0-5 cm with I (p = 0.022; R² = 0.174); volume 2-5 cm with I (p = 0.017; R² = 0.188); volume 2-5 cm with E (p = 0.043; R² = 0.138); MCA1 with I (p = 0.0004; R² = 0.366); MCA1 with E (p = 0.005; R² = 0.244); MCA2 with I (p = 0.001; R² = 0.407); and MCA2 with E (p = 0.002; R² = 0.29). Conclusions: There was correlation between the variables of CTCB, AR and CR in MB children with maxillary atresia.

STATE OF THE ART: 24 YEARS OF MOLECULAR ANALYSIS OF CYSTIC FIBROSIS IN HOSPITAL INFANTIL JOANA DE GUSMÃO, FLORIANÓPOLIS SANTA CATARINA

Louise Lapagesse de Camargo Pinto; Norberto Ludwig Neto; Eduardo Picciantini; Monica Lisboa Chung Waihs; Luiz Roberto Agea Catoio; Vanessa Platt; Hospital Infantil Joana de Gusmão

Objective: Describe the genetics of cystic fibrosis in a center of reference in the field and describe the flowchart of the molecular research of the CFTR gene. Material: 137 medical records were reviewed at Hospital Infantil Joana de Gusmão. Methodologies: cross-sectional study with 137 children of 0–15 years. Project approved by the Ethics Committee of the Hospital Infantil Joana de Gusmão. After a positive screening and the confirmation of the diagnostic with sweat electrolyte test, research was done on the DF508 mutation or on other 3 mutations (DeltaDF508, R553X, N1303 K). If the results were negative, patients were screened for 36 other mutations. Results: 91/137 (67%) patients had at least one mutation identified and the most common in 34 (50%) was DF508 in one allele. In 52 patients, 2 mutations were identified and 30 were homozygous for DF508. The second most common mutation in 10 patients was G542X (7 heterozygous). However, in 30 (33%) patients no mutation were identified. Conclusion: cystic fibrosis is an autosomal recessive disorder that intrigues from the genetics point of view, due to its broad genetic heterogeneity with 2006 mutations described so far. The flowchart of the molecular analysis allowed for the identification of most genotypes of the patients. The most common mutation, as expected, was the DF508, but the increase in frequency of the G542X mutation in our sample draws attention. The next step will be the sequencing of the CFTR gene in patients without the genotype. With the advent of new therapies based on genotype, the identification of mutations should become part of the standard procedure for patient monitoring.

RESPIRATORY COLONIZATION AND NUTRITIONAL STATUS OF CHILDREN AND ADOLESCENTS IN THE CYSTIC FIBROSIS CLINIC FROM CLINICAL HOSPITAL-UFG

Marília da Silva Garrote (Hospital das Clínicas- Universidade Federal de Goiás); Lusmânia Daucaneco Camargo Costa (Hospital das Clínicas – Universidade Federal de Goiás); Mario da Silva Garrote Filho (Universidade Federal de Uberlândia); Warley Diego Francisco Duarte (Hospital das Clínicas – Universidade Federal de Goiás); Rafaela Fangaro Baragatti (Hospital das Clínicas – Universidade Federal de Goiás); Raquel Vídia Fernandes (Hospital das Clínicas – Universidade Federal de Goiás)

Objective: To evaluate the relationship between nutritional status and respiratory colonization in children and adolescents with cystic fibrosis. Material: Children and adolescents treated in a specialized clinic at the Clinical Hospital- Federal University of Goias, between January – November 2015. Methodology: This cross-sectional study evaluated medical records of 40 patients whose information was collected through structured form containing demographic, anthropometric data and results from respiratory secretion culture (sputum of cough swab). BMI percentile (pBMI) was used for nutritional classification, considering malnourished those in the lower 10th percentil. Results: Thirty four patients (20 female, 14 male) who had at least one respiratory secretion culture held in 2015 recorded in the chart were included. Most patients (n = 20, 58.8%) were female, the mean age was 10.2 ± 5.3y (mean age at diagnosis 1.3 ± 4.2y); Most patients (n = 31, 91.1%) had some positive culture, such as Staphylococcus aureus (n = 22, 64.7%) Pseudomonas Aeruginosa (Pa) (n = 18, 50%), mucoid Pa (n = 1, 2,9%). Regarding to nutritional status, 12 patients, (35.2%) had pBMI>50th, according to CF Foundation recommendation, and 14.7% were overweight (pBMI>75th). On the other side, 12 patients (35.2%) were malnourished and 17.6% had nutritional risk (pBMI 10-25th). The proportion of malnourished who were Pa positive was high (83.3%), and the risk of those patients was 2.3 times higher than those non-malnourished of having that colonization (IC 1.2–4.2). Conclusion: Most patients in this study did not attend the CF recommendation regarding nutritional status and we observed higher prevalence of positive culture of Pseudomonas in malnourished patients.

ANALYSIS OF ASTHMATIC ADOLESCENTS BY GINA 2015

Andréa Lebreiro Guimarães Venerable: Ingrid dos Anjos Teixeira; Ana Luiza da Costa Barbosa; Rômulo Piloni Parreira; Bruna Santos Pacheco; Glauca Macedo de Lima; Escola de Medicina Souza Marques (FTESTM)

Assess control, risk factors, current treatment and quality of life. Study carried out in a Municipal Hospital in Rio de Janeiro, in 100 adolescents (12–19) randomly selected, taking controller treatment for 6 months. Was collected information about age, sex, adherence, inhaler technique(IT), peak expiratory flow(PEF) and control by ACT. Self-evaluation of quality of life was assessed by a personal note, from one to ten, with 1 being the worst and 10 the best quality of life. It was also applied a questionnaire on Quality of Life in Asthma in Young with Standardized Activities – Juniper (1992) – brazilian version. It was considered bad quality of life a personal note below 4 and the sum of answers in which they felt uncomfortable. Under the approval of the Ethics Committee in Research of the Municipal Health and Civil Defense. The responsible signed consent forms authorizing their participation. Cross-sectional, descriptive and analytical study. Data were collected and analyzed in Excel. The relative risk and chi-square test was assessed with 95% confidence interval. There was 64 males (mean age 14.11, median 14, SD 1.92) and 36 females (mean age 14.64, median 14, SD 2.21). ACT uncontrolled (20), well-controlled asthma(80). Adherence in front of the responsible(89), when distant(53). Incorrect IT(89). PEF < 2.21). ACT uncontrolled (20), well-controled asthma(80). Adherence in front of the responsible(89), when distant(53). Incorrect IT(89). PEF < 2.21). ACT uncontrolled (20), well-controled asthma(80). Adherence in front of the responsible(89), when distant(53). Incorrect IT(89).
step 5 ($X^2 = 13.20$ and $p < 0.005$). In front of the responsible they referred more adherence, which could indicate that they not feel comfortable to be honest near them. Incorrect inhaler technique should be identified at each visit. Quality of life by literature instrument was able to identify those in step 5. Despite being identified 80 well-controlled, 30 had low lung function.

125 PHYSIOTHERAPY AND NONINVASIVE RESPIRATORY MANAGEMENT AS ADJUVANT TO PREVENT EARLY TRACHEOSTOMY IN SPINAL MUSCULAR ATROPHY TYPE I

Paulo André Freire Magalhães (Programa de Pós-graduação em Saúde Materno-Infantil, Instituto de Medicina Integra Prof. Fernando Figueira); Ana Carolina Gusmão d’Amorim (Real Hospital Português de Beneficência em Pernambuco); Ana Patrícia Aquino Mendes (Real Hospital Português de Beneficência em Pernambuco); Maria Eveline Albuquerque Ramos (Real Hospital Português de Beneficência em Pernambuco); Márcia Beatriz Santos de Almeida (Associação de Assistência à Criança Deficiente – AACD); Maria do Carmo Menezes Bezerra Duarte (Programa de Pós-graduação em Saúde Materno-Infantil, Instituto de Medicina Integra Prof. Fernando Figueira)

Objective: Spinal atrophy type 1 (SMA) is an autosomal recessive genetic disease which promotes cell death of motor neurons located in the ventral horn of the spinal cord and brainstem nuclei engines. Early morbidity and mortality in this group is associated with bulbar dysfunction and respiratory failure requiring hospitalization and artificial ventilation. A large proportion of this population is considered difficult to wean from mechanical ventilation. Tracheostomy is considered treatment of choice for children with SMA 1.

However, recent studies have been suggested noninvasive respiratory management as adjuvant to prevent early tracheostomy. The aim of the present study was to promulgate awareness of maintaining a patient with SMA 1 without the use of invasive ventilation and with a personalized physiotherapy protocol, thereby providing better quality of life and integration with family members. Case description: a case study of a minor diagnosed with SMA 1 who, at the age of twelve months, underwent invasive mechanical ventilation (IMV) for 76 days, with successful weaning after application of a respiratory physiotherapy protocol, including use of mechanically assisted coughing and non-invasive ventilation (NIV).

Discussion: despite the difficulties and complications observed, the assistance proposed achieved the objective of removal of IMV and transfer to home care using non-invasive ventilation. IMV via tracheostomy is the treatment of choice in Brazil but families need to be informed of the irreversibility of the disease and the strategic possibilities of current therapies (IMV, NIV and palliative care) for management of a child with severe SMA 1.

126 RARE CFTR GENE MUTATIONS: IS IT CF OR NOT?

Fabíola Villac Adle (Pediatric Pulmonology Division – Instituto da Criança, Hospital das Clínicas – Medical School of USP); Salmo Raskin (Group for Advanced Molecular Investigation, Graduate Program in Health Sciences, School of Medicine)

Objective: To describe 3 patients with rare CFTR gene mutations and correlate them with the clinical presentation. Case reports: FAMILY A) Male siblings born from a CF mother [F508del/c.3140-26A>G mutations; pancreatic sufficient (PS), diagnosis at 34 years]. One is 7 y.o., had an increased IRT at newborn screening, normal/borderline sweat tests. CFTR full gene analyses showed F508del/p.R851L mutations. Nutritional status is normal, he is PS and has mild upper airway infections. His brother is 4 y.o., had an increased IRT, normal sweat test, but F508del/p.R851L mutations were detected. He had a gastroesophageal reflux (GER) in the first year of life and has mild upper airway infections. Nutritional status is normal and he is PS. FAMILY B) A 7 y.o. male, had an increased IRT and normal/borderline sweat tests. In his first year he had GER and electrolytes abnormalities. Two sweat tests performed at 2 y.o. were abnormal. CFTR gene analysis detected the R117H mutation (intron 8 polyT alleles: 7T/9T) and a deletion from exon 17a through 18. The R117H mutation was found in the mother and the deletion in the father. The only respiratory manifestation he has is recurrent sinusitis. Nutritional status is normal and he is PS.

Discussion: the p.R851L mutation, also known as c.2552G>T, localized in exon 15 of the CFTR gene, was previously detected in the father of 2 CF patients who died and were probably homozygous for the R851L mutation. The deletion in the CFTR gene from exon 17a to 18 has been seldom reported, either associated with mild or severe disease. R117H is a type IV CFTR mutation and when in compound heterozygosity with other mutations present a wide phenotype variability. Rare mutations in the CFTR gene require strict follow up, monitoring digestive and respiratory symptoms and nutritional status. The clinical picture of the 2 siblings support the diagnosis of CFTR related metabolic syndrome while the other patient manifestations support the diagnosis of atypical CF.
ENDOBRONCHIAL APPLICATION OF FACTOR VIIA IN A PEDIATRIC PATIENT WITH DIFFUSE ALVEOLAR HEMORRAGE (DAH) AND SYSTEMIC LUPUS ERYTHEMATOSUS (SLE)

Velasquez Mendez, Monica (Hospital San Vicente Foundation); Johana Hernandez Zapata (Universidad de Antioquia); Olga Lucia Morales Munera (Universidad de Antioquia y Hospital San Vicente Fundacion)

Objective: To report administration of recombinant activated factor VII (rFactor VIIa) by endobronchial bronchoscopy for treatment of DAH in pediatric patients with SLE without response to usual medical treatment.

Methodology: Case report. Results: Male teenager (13 years), who was high, showing that spirometry is a valid method for assessing pulmonary function in very young (preschool) children. The high success rate in our sample might be attributable to the use of an incentive and to the fact that the tests were performed by professionals specializing in pediatrics.
by several specialties with close monitoring. Conclusions: In countries with high prevalence of tuberculosis, it should be considered as a primary differential diagnosis specially in immunocompromised patients, but it is essential to keep in mind that other opportunistic pathogens may be concomitantly present and being cause of poor response to treatment.

132 SURFACANT DEFICIENCY TYPE 2 RELATED TO THE MUTATION OF THE GENE SFTPC IN PEDIATRIC PATIENT
Claudia Renato Alves Alcure; Lais Fraga Pereira; Camila Barros Braga Miranda; Mariana Pandolfi Pina; Flávia Miguel Vervloet; Vitor Earl Vervloet; Hospital Infantil Nossa Senhora da Glória

Objective: Report a case of respiratory failure associated with mutation of the gene SFTPC. Case Report: SRMP, male, 2 months, admitted to the service in Feb/2014 with dyspnea that progressed to respiratory failure and mechanical ventilation (MV). No response to clinical treatment. Conducted Tomography (CT) showed chest with small airway commitment, peribronchial consolidation. Hypothesized broncholitisi obliterans. Conducted 3 cycles of pulse therapy, tolerated suspension of MV. Discharged in Dec/2014, with home oxygen therapy. He returned in Apr/2015 with pneumonia and need for intubation and MV. Required progressive increases in MV ventem and evolved with episodes of pneumothorax. New lung biopsy suggesting proteinaceous material intra-alveolar and blood sent for molecular research. Made in bronchoalveolar lavage Aug/2015 with milky material output and the possibility of reducing the MV parameter. Another lavage in a month showed less milky material and impossibility of reducing MV parameter. The patient developed recurrent pneumothorax, infectious worsening, hemodynamic instability and death in Nov/2015. In Dec/2015, panel showed molecular presence of heterozygosity in SFTPC gene associated with dysfunction of the pulmonary surfactant metabolism type 2, with autosomal dominant inheritance. Discussion: Intersitial lung diseases are rare in pediatrics and may be caused by specific inborn errors of surfactant metabolism. Deficiency and mutations in SFTPC have been associated with interstitial lung disease. The mutation SFTPC occurs threonine substitution for isoleucine at codon 73. Age of onset and severity of lung disease varies, from early death to pulmonary fibrosis at the fifth/sixth decade of life. There are more than 35 mutations in the gene SFTPC, however, it is not clear which of these mutations determine the signs and symptoms. Those altered proteins can trigger cell damage and death. Lung transplantation is only definitive treatment option.

133 GENETIC VARIANTS ASSOCIATED WITH LUNG FUNCTION IN ALLERGIC ASTHMA
Fernando Augusto de Lima Marson (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Carmen Silvia Bertazzo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Maria Angela Gonçalves de Oliveira Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Adyélia Aparecida Dalbo Contrera Toro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas); Tânia Kawasaki de Araújo (Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas) José Dirceu Ribeiro (Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas)

Objectives: To verify the association of genes related to inflammatory response with clinical markers of spirometry. Material: Included 221 asthma allergic (AA) patients. Methods: Spirometry was performed based on international standards (European Respiratory Society). For spirometry were considered: FVC (forced vital capacity), FEV1/FVC and forced expiratory flow between 25–75 percent of FVC (FEF25-75%). 256 polymorphisms (pol) on 125 genes were considered for association study. For genetic analysis, OpenArrayTM platform (LifeTechnologies®) was used. SPSS vs 22.0 performed the statistical analysis. alpha = 0.05. Results: The pol analyzed were associated with spirometry data before bronchodilation. The values were compared in percentage of predicted. Each spirometry marker shows association with particular genes. FVC was associated with 15 pol in 14 genes [CDH1, FCR1A (2 pol), GIPC3, IKZF3, IL18RAP, IL23R, IL33, IL5R, IRAK3, IRF4, NR3C1, SOCS5, TBX2AR and TLR4]. FEV1 was associated with 7 pol in 6 genes [CDH1 (2 pol), IL12B, IL33, IL5R, NR3C1 and SOCS5]. FEV1/FVC was associated with 23 pol in 13 genes [CA10, HRH4, IL12B (5 pol), IL17F, IL18, IL33, IL4R (2 pol), IRF5, ITGB3 (2 pol), NAP53, RNASE3 (three pol), RUNX1 and TGBF1]. FEF25-75% was associated with 19 pol in 14 genes [CA10, HRH4, IL12B (4 pol), IL17A, IL18, IL4R (2 pol), IL5R, IRF5, ITGB3 (2 pol) NFKBIA, NAP53, RNASE3 and RUNX1]. Conclusion: The AA severity assessed by spirometry features is determined by multiple genes and their variants. The genetic pattern associated with the gravity is extremely heterogeneous, showing variation among AA patients and among the markers evaluated by spirometry. To understand this complex variability many studies are necessary.

134 CASE REPORT: MEDIASTINAL CYST LEADING TO BRUISES ON LOWER MEMBERS BY DESCENDING AORTA COMPRESSION
Eduardo Augusto Caldeira Storti; Renan Augusto Pereira; Hospital Pequeno Principe

Objective: To report a case of a 4-year-old preschooler with an atypical clinical presentation of mediastinal cyst. Case Report: PJ, 4 years-old, presented with hematomas on lower limbs for approximately 4 months, associated with daily fever (38.5 °C, once a day) in the last month. In a routine chest radiography, it was observed a 3-cm-nodule compressing the mediastinal surface of the left lung. Chest computed tomography and magnetic resonance showed an oval and well-demarcated cyst in left posterior mediastinum, between T6 and T7 vertebrae, with 8.5 cm3, compressing 30% of descending aorta, and suggesting bronchogenic cyst. Blood screening tests for neoplasia were negative, and the transthoracic ecocardiography, abdominal ultrasound and tuberculosits tests were negative either. Currently, the patient awaits the surgical extraction of the mediastinal lesion and the anatomicopathologic study, to confirm the etiology. Discussion: In children and infants, neurogenic tumors are the most commonly occurring mediastinal tumors; bronchogenic and other rare types of cysts comprise 10 to 18% of all mediastinal masses identified. Constitutional symptoms, such as weight loss, fever, malaise, and vague chest pain can occur in the pediatric population. Persistent cough, dyspnea, stridor and other respiratory tract symptoms predominate in pediatric patients, although signs of compression of adjacent organs, such as esophagus, and less likely, vascular structures (like in our case), may be present. Bronchogenic cysts are asymptomatic in as many as 20 to 30% of children. When symptomatic, children most commonly present with vague respiratory problems or dysphagia. Surgical resection of all symptomatic bronchogenic cysts is recommended; thoracoscopic resection has major advantages compared to classic thoracotomy. Palliative procedures, such as aspiration, may be considered in cases in which patients are symptomatic and the situation for complete resection is not optimal.

135 BRONCHIOLITIS OBLITERANS POST-BONE MARROW TRANSPLANTATION
Jaqueline Paso Blans Carmelos; Talitha Di Marina Chacon Belotti; Victor Gottardello Zechin (Instituto Oncológico Pediátrico – Universidade Federal de São Paulo); Sonia Mayumi Chiba; Beatriz Neuhaus Barbisan; Clovis Eduardo Tadeu Gomes; Universidade Federal de São Paulo

Pediatric Pulmonology
Objective: Describe a case of Bronchiolitis Obliterans (BO) post Allogeneic Related Bone Marrow Transplantation (MRD-BMT). Case report: A 10.5 year old male patient was diagnosed with Acute Myeloid Leukemia. At age 11 he received MRD-BMT due to induction failure (Universidade Federal de São Paulo). Seven months after the transplant, Prednisolone and Cyclosporine were suspended. One month later (Oct 2014) the patient presented cutaneous and ocular manifestation of chronic Graft Versus Host Disease (cGVHD) and some weeks later, dry cough. Percent-of-predicted FEV1 was 114 in July and 60 in November. Computed Tomography (CT) showed nodular opacities, “tree in bud” pattern and bronchial wall thickening in lingula and middle lobe. Prednisolone (1 mg/kg/d), Cyclosporine and Azithromycin were reintroduced. In Dec 2014 his lung function test improved suggesting response to treatment. It showed an improvement with bronchodilator in FEV1% of 75 to 82 and was introduced formoterol/budesonide. On Jan 2015, however, patient reported dyspnoea at moderate efforts and SpO2 fell to 93% post exercise. FEV1% decreased to 48 and in 2 weeks to 36%. CT remained unchanged. Methylprednisolone pulse therapy (10 mg/kg/d) for 3 days, Imatinib and inhaled Cyclosporine were instituted and patient improved of dyspnoea and cough after. On 2/17/2015, the child had a sudden episode of breathlessness during the night and died before the arrival of medical aid. Discussion: Incidence of BO post BMT ranges from 2 to 26%. It affects 10% of patients that develop cGVHD. Diagnosis is suggested by FEV1% < 80, once infection has been excluded. In general, change in spirometry comes first than classical symptoms. Treatment includes steroids and Cyclosporine, but the answer is usually poor. It is a rapidly progressive disease; we stress the strict need of monitoring pulmonary function after BMT to allow early treatment and reduce mortality. In this case death occurred by unknown causes, probably unrelated to BO.

136 CLINICAL PROFILE OF SICKLE CELL DISEASE CHILDREN WITH COMMUNITY ACQUIRED PNEUMONIA (CAP) IN A REFERENCE SERVICE

Paula Rosalina Oliveira de Radenauer Rayaiba; Clemas Couto Santanna; Maria de Fátima Banzuni Pombo March; Sidney Ferreira; Ana Alice Amaral Biaupina Parente; Priscilla Aguiar de Araújo; Instituto de Puericultura e Pediatria Martaça Gesteira

Objective: Evaluate the main clinical profile of sickle cell disease children with CAP. Methodology: Transversal retrospective descriptive study with children aged 0–12 years with sickle cell disease and CAP admitted at IPPMG from 2004 to 2011. Standardized questionnaires were filled with medical records from admission of CAP: clinical and epidemiological history, signs, symptoms, risk factors, radiological and bacteriological patterns, antibiotic therapy and clinical outcome. Results: 475 children were identified with CAP, 76 (16,10%) had sickle cell disease. The average age of sickle cell disease children with CAP was 8 years and 3 months. 46 (60,53%) were male. 2 (2,63%) children went to nursery school and only 30 (39,47%) had actualized vaccine card. No child had asthma. 43 (56,58%) were previously admitted for pneumonia. 49 (64,47%) children had cough and 62 (81,58%) fever, which lasted 1,59 days. Breathlessness was reported in 36 (47,37%) patients. Tachypnea was found in 43 cases (56,57%). Children’s general condition was good in 42 patients (55,26%). Alveolar lobar consolidation was confirmed in 48 cases (63,16%) while interstitial infiltrators in 7 (9,21%). Pleural effusion was found in 14 patients (18,42%). G penicillin was prescribed for 44 children (57,89%) and second generation cephalosporins for 36 (47,37%). Blood culture was collected in 67 (88,16%) patients, of which 3 (3,95%) were positive. Pleural samples were collected in 10 of 14 patients, of which 8 went to bacteriological analyses (1/8 positive). Discussion: There was a male predominance and nearly half of the children had previous history of CAP. The vaccine coverage was inadequate. The most common radiological finding was alveolar lobar consolidation. The main pulmonary complication was pleural effusion. No child had severe clinical outcome. G penicillin was the main prescribed drug and all sickle cell disease patients had satisfactory resolution of CAP.
place. After 8 months, his weight increased weight and LF improved (BMI/A = 20.2, H/A = 0.2, FEV1 = 33.2%). However, although the diet prescription was adequate in energy, after 1 year, he returned to FN (BMI/A = 0.2) and decreased FP (FEV1 = 32.9%), suggesting difficulties of adherence to nutritional intervention. Discussion: The nutritional status of CF children and adolescents undergoing GTF improved 1 year after the procedure, although not constantly. The nutritional status reflected in LF improvement. It is suggested earlier intervention and better monitoring to ensure better nutritional status and clinical outcomes.

139 TUBERCULOMA IN CHILDREN: A CASE REPORT
Priscilla Aguiar de Araujo; Bruna de Paulo Santana Rafaela Baroni Aurillo; Ana Alice Amaral Ibiapina Parente; Maria de Fátima Bazhuni Pombo March; Clemax Couto Sant’Anna; Instituto de Puericultura e Pediatria Martágao Gesteira (IPPMG/UFRJ).

Objective: Present a case of child tuberculosis, rare form of pulmonary tuberculosis (PT) that simulates tumor. Case Report: Girl, 2 years old, with fever, vomiting, diarrhea and sweating. Went to the emergency, where she performed chest X-ray: consolidation in middle 1/3 of the right hemithorax (RHT), and blood count: leukocytosis with deviation. She was admitted and used various antibiotics. Chest CT: mass in the upper mediastinum right (2.8 x 2 cm), with calcification. Nuclear magnetic resonance: consolidation in the right upper lobe, subpleural nodular image in the middle lobe and right mediastinal lymph nodes with central necrosis. Tuberculin skin test (TST): 8 mm. Gastric lavage: negative AFB. Without BCG scar and no history of contact with a tuberculosis patient. Vesicular murmurs diminished in upper third of RHT. Treated with HRZ scheme during 6 months, with clinical and slight radiological improvement. Discussion: The PT is caused by Mycobacterium tuberculosis. One of its forms of presentation is the tuberculosisomast, consisting of well-defined mass or lump simulating tumor. Normally is unique, measuring about 1 to 10 cm in diameter, located in the upper lobe and may contain calcifications or cavitations. It is 5–24% of the causes of solitary pulmonary nodules. Usually presents benign evolution, being considered healed mechanism of the PT. Neoplasias, histoplasmosis and coccidiodomycosis are its differential diagnoses. Although rare, it is more common in adult-type tuberculosis. The diagnosis is clinical (symptoms similar to PT) and radiological. The anatomical and pathological investigation, by puncture and aspiration or open thoracotomy, may be required in special cases.

140 NEWBORN SCREENING: PREVALENCE OF CYSTIC FIBROSIS AND F508del MUTATION FREQUENCY IN THE STATE OF GOIÁS
Lasmaia Damaceno Camargo Costa; Lian Padovez; Cualheta (HC-UFG); Alaine Rodrigues Belo (HC-UFG); Virginia Auxiliadora Freitas de Castro (APAE-Anápolis); Bruno Bento Rodrigues (APAE-Anápolis); Flaviane Marcília Pedatella (APAE-Anápolis).

Objective: To describe the results of newborn screening for cystic fibrosis in the state of Goiás, its frequency and the F508del mutation in this population.
Methods: This cross-sectional study analyzed the number of newborn screening tests, abnormal sweat testing, genetic mutation and the average age of the infants at the time of first the altered sweat test in the reference center for neonatal screening in the state of Goiás. Results: Neonatal screening for cystic fibrosis (CF) is held in Goiás since November / 2009, and there were currently 26 CF patients in follow-up at the APAE-Anápolis Center of CF. Data of the period 2011-2014 were recovered, and 299,719 neonatal screening tests were performed during that period. There were 1096 positive IRT (IRR > 70 ng/dl) in the first test, 23 in the second test and 18 had been confirmed thru the Sweat Test (Chloro> 60 mEq/L), leading to a prevalence of 1: 18.512 FC in this population. The F508del mutation was studied in 14 patients, positive in 10 (71.4%) patients. 4 being homozygous. The average age at diagnosis (performing the sweat test) was 48.23 days. Conclusion: The neonatal screening in the state of Goiás enabled early diagnosis of cystic fibrosis, in children having less than 60 days of life where the F508del mutation was present in more than half of the patients.

141 DIAPHRAGMATIC EVENTRATION RIGHT MIMICKING PNEUMONIA
Cleber Eduardo Franchin dos Santos (Faculdade de Medicina Ingo – Maringá –PR); Nathalia Ingrid Boer (Faculdade de Medicina Uninag); Livia Fonseca Calheiro (Faculdade de Medicina Uninag).

Objective: report a case of a boy 3 months old with eventration diaphragmatic right mimicking a pneumonia lobar. Case: A male patient, 3 months old, that was admitted into an emergency service reporting runny nose and dry cough. He presented CBC (normal), PCR 1.31, and chest X-ray, that revealed elevated right hemidiaphragm. The pediatrician treated the case as bacterial pneumonia, and started the treatment with intravenous ceftriaxone. On the fourth day, another X-ray was performed without any change. The patient was discharged with orally antibiotic to complete seven days of medication. On the seventh day, he was evaluated by a pediatric pulmonologist, presenting a good evolution. It was requested a new chest X-ray, and the image remained similar to previous exams. The diagnosis of a diaphragmatic eventration was suspected, and a CT scan requested. This exam confirmed the hypothesis of right diaphragmatic eventration. He is been monitoring and was referred for evaluation with pediatric surgeon. Discussion: The clinical manifestations, may vary from asymptomatic patients, including diagnosis can be realized only in adulthood, to neonates that may have acute respiratory distress at birth. In adults, the most common clinical presentations include recurrent respiratory infection, dyspepsia, dyspnea and chest pain. Most diagnoses are made by chest X-ray, and confirmed with fluoroscopy and CT if necessary. In some cases it is difficult to distinguish diaphragmatic eventration from congenital diaphragmatic hernia. The most reliable test is the fluoroscopy, which reveals a functional image of the paradoxical movement of the affected side. For patients who are asymptomatic or without surgery indication, the treatment is conservative. For symptomatic cases, the treatment of choice is surgery (placature of the diaphragm). Respiratory complications are more common in children than in adults, and the surgery is done to restore the volume of the lung parenchyma.

142 FOLLOW-UP OF CHILDREN CONTACTS OF TUBERCULOSIS IN A PRIMARY HEALTH CENTER IN RIO DE JANEIRO, BRAZIL
Paula do Nascimento Maia; Clemax Couto Sant’Anna; Maria de Fátima Bazhuni Pombo March; Paula Andrea Vaca Gonzalez; Bruna de Paulo Santana; Paula Rosalina Oliveira de Rademaker Itagiba; Instituto de Puericultura e Pediatria Martágao Gesteira/ UFRJ.

Objective: Evaluate clinical features of tuberculosis (TB) contacts who started isoniazide preventive therapy (IPT). Material and Methodology: Retrospective longitudinal study of children and adolescents TB contacts in a health center of Rio de Janeiro, between 2006 and 2014. They all were submitted to tuberculin skin test (TST) and chest X-ray and were evaluated by Guidlines of the Ministry of Health – Brazil (MOH) and classified as: active TB, latent infection of TB (LTBI) and exposed. Afterwards, they had their follow-ups analyzed. Results: 245 contacts were assessed, of which 227 were included in the study. Of these, 158 (69.6%) had LTBI, 13 (5.7%) had active TB and 56 (24.7%) were exposed. Of the 158 with LTBI, 157 (99.4%) received IPT of which 154 (98.1%) completed treatment. 3 (1.9%) withdrew, and 2 had no informations. Twelve (92.3%) patients with active TB were submitted to TB treatment recommended by MOH and 1 (7.7%) abandoned. Of the 56
exposed, 25 (44.6%) were discharged and 31 (55.4%) abandoned follow-up (out of sight). Conclusion: The clinical and epidemiological assessment, associated to TST and chest X-ray, allows the diagnosis and the proper conduct in childhood. The treatment of LTBI prevents illness and can diagnose new cases of TB disease. Patients with LTBI and TB disease showed good adherence. However, there were large percentage of dropout among exposed patients, emphasizing the importance of good reception and appropriate follow-up among TB contacts in a health center.

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WHEEZING IN INFANTS: EPIDEMIOLOGY, INVESTIGATION AND RESPONSE TO TREATMENT

Raquel Mascarenhas Freitas; Thiara Maria Gomes da Silva; Edison Tapina Braga; Carlo Roberto Bazzo; Rai Andre Silva Watanabe; Clovis Eduardo Tadeu Gomes; Universidade Federal de São Paulo

Purpose: To evaluate the epidemiological aspects, risk factors and treatment of wheezing in infants. Methods: We reviewed 139 medical records of patients followed at Federal University of São Paulo in the wheezing infant clinic of Pediatric Pneumology, from August 2013 to March 2015. Results: We studied patients from 5 to 44 months with a male prevalence (60.4%). Birth weight: <1000g, 4 (3%); between 1000 and 2000g, 13 (9%); between 2000 and 3000g, 45 (32%). Frequency at daycare: 43 (31%). Parental atopy: 43 (31%). First episode of wheeze: 0 to 2 months, 54 (39%); 3 to 5 months, 43 (31%); 6 to 8 months, 26 (19%); 9 to 11 months, 4 (3%). Triggering factors of the crisis: 51 (37%) attached to the upper airway infections, 41 (29%) to the weather and 26 (19%) to both of them. Frequency of the crisis: weekly 26 (19%); each fifteen days, 27 (19%); monthly, 50 (36%); each 2 months, 17 (12%). Related diagnoses: asthma 58 (41.7%); asthma with gastroesophageal reflux 19 (13.6%); post-viral hyperreactivity, 29 (20.8%); post-viral hyperreactivity associated with gastroesophageal reflux, 12 (8.6%); bronchopulmonary dysplasia, 7 (5%). Suggested therapy: inhaled corticosteroids, 73 (52.5%); inhaled corticosteroids associated with domperidone and ranitidine, 31 (22.3%); inhaled corticosteroids and domperidone, 8 (5.8%). Improvement of symptoms with the suggested therapy, according to those responsible for the child, after three months of first consultation: good improvement, 59 (43%); little or some improvement, 27 (19%); no improvement 8 (5%). Conclusions: Recurrent wheezing in infants is a heterogeneous condition usually related to a bronchial hyperresponsiveness, but may be present in any situation that leads to a narrowing of the airways. A careful clinical evaluation is important to infer the possibility of asthma and exclude other diseases that require specific approaches, this way to determine the appropriate therapy.

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CISTIC FIBROSES IN CHILDREN AND ADOLESCENTS IN FORTALEZA – CE: A EPIDEMIOLOGY STUDY AND COMORBIDITIES

Cláudia de Castro e Silva (Universidade Federal do Ceará ( UFC) e Hospital Infantil Albert Sabin(HIAS)); Hildenia Baltazar Ribeiro Nogueira (Universidade de Fortaleza/UNIFOR) e Hospital Infantil Alberto Sabin (HIAS); Amanda Vale Catunda(1); Débora Ponteiró Giñoni(1); Patricia Sampaio Padilha; Vanessa Guerreiro Soares (1); I. Universidade de Fortaleza (UNIFOR)

Objectives: Understand epidemiological profile of patients with Cystic Fibrosis (CF); Identify the patient clinical status and main comorbidities at in specialized clinic. Procedure: a descriptive, quantitative study. Realized through retrospective analysis of clinical data of 55 patients of the Children’s Hospital Albert Sabin, Fortaleza CE. The data acquisition and analysis was between August and December 2015. The study included patients diagnosed with the CF in follow-up at the clinic. Results: The patients age range between 8 week to 15 years,average age being 4 years,40 weeks; The average age range of admission was 2 years and 32 weeks. 65.45% (36) of the patients live in cities far from the hospital; 52.73% (29) are female; The average BMI is 15.25 kg/m2; 21.8% (12) had a z-score lower than – 3; 21.8% (12) had a z-score ranging between – 3/-2; 36.6% (20) had a z-score ranging between – 2/0. The comorbidities were: 27.27% with allergy to cow’s milk protein; and 5.45% celiac disease. Minor percentage of tuberculosis, asthma. Between symptoms, the productive cough was in 78.1%; 72.7% were below the proper weight; 56.4% had recurrent airway infections; 47.3% had diarrhea with mucus; 41.8% for vomit and pneumonia; 27.3% abdominal distension and 9% symptoms of liver disease. Conclusion: The association of comorbidities such as celiac disease and allergy to cow’s milk protein can be present in patients with cystic fibrosis and should therefore be part of the routine investigation. The patients that live far from the clinic end up arriving there too late. The low weight and recurrent respiratory infections symptoms reaffirm the delayed arrival of the patients, once it is related to lack of good prognosis. This scenario could be changed with early diagnosis and treatment. Thus this study to point the need for revaluation of forward flow for the newborn screening results in a better survival for children with CF accompanied by the reference service.

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A MILD PANCREATIC SUFFICIENT CF-PHENOTYPE ON TWO UNRELATED PATIENTS WITH THE I148N:ST MUTATION

Luísa Mesquita Nunes (Centro de Pesquisa Experimental, Hospital Israelita Albert Einstein); Vivian D. T. Newiandowski (D iagnóstico das Américas S. A); Nelson Gaburo (D iagnóstico das Américas S.A); Guilherme Lopes Yamamoto (Instituto da Criança, Universidade de São Paulo); Débora Romeu Bertola (Instituto da Criança, Universidade de São Paulo); Luiz Vicente Ribeiro F. da Silva Filho (Centro de Pesquisa Experimental, Hospital Israelita Albert Einstein; Instituto da Criança, Universidade de São Paulo)

Objectives: To describe two unrelated patients presenting with mild and pancreatic sufficient CF and the genotype c.[443T>A;1210-12[5] (I1521_1523delCTT) (I148N:ST/F508del). Case report: Patient L.H.J., female, 17 years old, was referred to our center in January/2013 with recurrent nasal polyps and two borderline sweat tests (59,4 and 54,4mEq/L). The patient was enrolled for follow up with a suspicion of atypical CF. At the time of diagnosis she had no pulmonary or GI tract symptoms; adequate height and weight for age and normal lung function (FEV1 102%). Orpharyngeal cultures were negative until August/2014 when MSSA was identified. Since diagnosis she developed sporadic cough and chest pain with no need for oral or intravenous antibiotics and remains pancreatic sufficient. Her latest lung function is normal with a decline in FEV1 to 86%. A chest CT was performed in November/2015 showing bronchial wall thickening and few dilated bronchi with inspissated mucus. E.N.C., female, 5 years old, was referred in July/2011, at 15 months old after 2 prolonged hospital stays for wheezing, 3 episodes of dehydration and failure to thrive. Two sweat tests confirmed CF (76,1 and 84,3mEq/L) and she started follow up at our Center. She showed no signs of pancreatic insufficiency and was referred to nutritional counseling. Since the start of follow up she has had few pulmonary exacerbations treated with oral antibiotics; remains pancreatic sufficient, with adequate weight and height for age; is colonized with MSSA and is not receiving any chronic medications. Her latest Shwachman-Kulczycki score is 90, and a chest CT in November/2013 showed bronchial wall thickening and inspissated mucus. Discussion: The I148N mutation was described on the CFTR mutation database in a Brazilian patient with no phenotypic information. It is possible that, similar to R117H, I148N has varying clinical consequence and can be CF-causing when in cis with a 5T polymorphism, causing mostly mild disease.

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MYCOBACTERIUM TUBERCULOSIS INFECTION IN CHILDREN WITH CYSTIC FIBROSIS. REPORT OF FOUR CASES

Pediatric Pulmonology
Mycobacterium tuberculosis (MTB) infection is rarely seen in cystic fibrosis (CF) patients. Most mycobacterial infections are caused by non-tuberculous mycobacteria. We report four CF patients screened for MTB. Results: 29 patients followed at the outpatient clinic of CF in 2015, 4 with pulmonary TB contact. Case 1: 4-year-old boy admitted to hospital for respiratory exacerbation, referred household TB contact. Tuberculin skin test (TST) was positive. Sputum culture: S. aureus, positive acid-fast bacilli smear. MTB Chest tomography (CT) scan showed mediastinal lymph nodes and bilateral centrilobular nodules. He received broad spectrum antibiotics plus isoniazid (INH), rifampicin (RIF), pyrazinamide (PZA), ethamubutol for 2 months, followed by 7 months INH and RIF. Case 2: 4-year-old patient, household pulmonary TB contact. Boy had failure to thrive for the last 3 months despite appropriate treatment. TST 10 mm, CT scan showed new rounded peripheral image. Sputum culture MTB negative. Treated with INH, RIF 6 months, plus PZA for first 2 months, with good therapeutic response. Case 3: 17-year-old patient consulted for respiratory exacerbation and hemoptysis. Contact with school classmate smear-positive TB. He presented TST conversion. Sputum culture negative for MTB, positive for Pseudomonas aeruginosa. After 2 weeks of intravenous antibiotic treatment he had clinical improvement. CT scan and pulmonary function tests unchanged. INH preventive treatment administered for 6 months. Case 4: 8-year-old boy, household TB contact. Clinical condition, chest X-ray and spirometric findings had no changes compared to previous status. TST, sputum smear and culture were negative. Received INH preventive therapy until contact screening was performed to his family and TST conversion was ruled out. Conclusions: TB diagnosis can be delayed due to preexisting pulmonary symptoms and radiological abnormalities in CF patients. Close tuberculosis exposure must be carefully evaluated.

147 EPIDEMIOLOGICAL EVALUATION AND FINAL DIAGNOSIS IN PEDIATRIC PATIENTS WITH REPORTED RECURRENT PNEUMONIA (RP)

Cátiva Codess Thome de Souza; Ana Paula Pereira da Silva; Daniel Lopes Aires; Universidade Estadual de Maringá

Objective: To assay data from patients treated at the pulmonary clinic complaining of RP, as well as the final diagnosis. Methods: A retrospective study based on the survey of the records of patients treated at the pulmonary clinic at University Hospital complaining of RP, from 02/01/15 to 10/30/15. This clinic assists about 25 patients/month. Results: There were 9 patients with a RP record. The mean age at the first diagnosis was 2 years and 4 months and 6 of them were females (66.7%). The average number reported of pneumonias was 4.6 episodes. 8 patients (88.9%) required previous hospitalization. 4 patients (44.4%) had comorbidities: anemia, Down syndrome, atopic dermatitis, obesity, esophageal atresia and encephalocoele surgically corrected. All patients presented reports of wheezing, only one child with swallowing disorder related. 6 children (66.7%) recorded parental health history of asthma – in the remainder, one sibling had rhinitis relate and the other one had asthma. Regarding the laboratory exams, there was no eosinophilia, and one medical record did not present this data. The PPD was performed in 4 children (44.4%), without positive results. Lastly, 8 patients (88.9%) had probable asthma diagnosis instituted during follow-up. Just one patient had a diagnosis of congenital agammaglobulinemia with pulmonary sequelae. This was the only one with radiological changes (bronchiectasis) and had no relatives of 1st degree with allergic respiratory diseases. Conclusions: The diagnosis of asthma was the most frequent in this group of patients. According to the literature, asthma may be an underlying cause that predispose to pneumonia and RP can be a risk factor for developing asthma. Also, sometimes the asthma attacks can be treated wrongly as pneumonia. Despite the young age, the reporting of wheezing and presence of positive parental history were determinant factors in diagnosing asthma.

148 MICROBIOLOGICAL PROFILE OF PATIENTS FOLLOWED UP IN A CF CARE CENTER IN PORTO ALEGRE, RIO GRANDE DO SUL, BRAZIL

Marina Melo Gonzalves; Débora Quiorato Fernandes; Luís Otávio S. Alves; Helena Teresa Mocelin; Gilberto Bueno Fischer; Universidade Federal de Ciências da Saúde de Porto Alegre

Objective: To describe the identification of microbiological profile from the respiratory tract of patients followed up in a CF care center. Methodology: Cross-sectional and prospective study. We analyzed microbiological identification data from sputum samples, swab of oropharynx and bronchoalveolar lavage of patients diagnosed by clinical or newborn screening (held since June/2012), collected in a routine service during the period 2001–2015. Bacterial resistance defined according to M100-S24 PSAST. Descriptive statistical analysis. Results: 1302 samples were included (32 with bronchoalveolar lavage) of 41 patients, median age 7.2 years (8m–19yo), 56% were male, 73% were white and none was black. The median age at the moment of the diagnosis was 8.5 m (1m–12 yo), only 34% after neonatal screening. Delta F508 mutation search was performed in 33%; 36% homozygotes, 27% heterozygotes and 30% negative. At least one germ was isolated in each patient. The frequency of germs in all samples was: S. aureus (SA) 35.8%; P. aeruginosa (PA) 16.6%; PA mucoid 10%; B. cepacia complex (BCC) 5%; Haemophilius sp. (Hm) 5%; S. marcensens (SM) 3%; Stenotrophomonas sp. (St); other pathogens (24 kinds) 16%; 6% negative. The frequency of germs per patient and the presence of resistance were: SA 88/33%; PA 63/21%; PA mucoid 27/22%; BCC 32/38%; Hm 54/9%; SM 34/43%; 51% of the patients had permanent colonization and 83% had resistant germs. Comparing the patients diagnosed by its screening/clinical status, the prevalence of germ per patient was: SA 93/85%; SM 57/33%; PA 42/74%; BCC 28/33%; Hm 28/51%; PA mucoid 71/33%; St 73/33%; oropharyngeal flora 100/96%; another 100/88%. Conclusion: All patients had at least one identified germ. S. aureus was the most common, regardless of the form of diagnosis and prevalent in chronic colonization, similar to what is described in the literature. The high rate of resistance and frequency of BCC should be considered in treatment and infection control measures.

149 CASE REPORT: INTRAPULMONARY SHUNT IN A POST BONE MARROW TRANSPLANTATION PATIENT WITH CONGENITAL DYSKERATOSIS

Débora Carla Chong e Silva; Camila Forestiero; Tatiane Guedes da Silva Scalante; Cristiane Secco Rosário; Carlos Antônio Riedi; Nelson Augusto Rosário Filho; Universidade Federal do Paraná

Objective: To report a case of intrapulmonary shunt in a post bone marrow transplantation patient with congenital dyskeratosis. Case Report: 12 year-old boy from Tocantins, Northern Brazil, underwent bone marrow transplantation due to bone marrow failure secondary to congenital dyskeratosis 4 years ago. Two years later, he began presenting progressive dyspnea and cough, now with minimal exertion. On physical exam, he was eupnic; room-air pulse oximetry was 83%; central cyanosis; diffusely diminished breath sounds; mild intercostal retractions; nail dystrophy; and digital clubbing. Initial investigation showed normal chest computerized tomography, lung volumes indicating restrictive pattern, lung biopsy with no signs of fibrosis or organizing pneumonia. Abdomen ultrasound showed hepatomegaly and liver enzymes were slightly elevated. Catheterization study found normal pulmonary artery pressure. The dyspnea kept worsening and we suspected of intrapulmonary shunt, which was confirmed by bubble echocardiography. Two months ago, he began supplementation with garlic capsules, 1g per day PO. Discussion: The most common manifestation of

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telomere syndromes is idiopathic pulmonary fibrosis and emphysema. The short telomere defect in these patients may manifest systemically as bone marrow failure and liver disease. In patients with congenital dyskeratosis, a telomere syndrome, and progressive dyspnea, the focus of the investigation is in post-bone marrow transplantation complications and the diagnosis intrapulmonary shunt is often forgotten.

150 RESULTADOS DE UNA ENCUESTA SOBRE MANEJO DE LA CRISIS ASMÁTICA EN NIÑOS EN LA PROVINCIA DE MENDOZA (REP. ARGENTINA). ¿HABLAMOS EL MISMO IDIOMA?

Fabian Alejandro Castracane; Programa Provincial de Asma Infantil de la Provincia de Mendoza.

En el año 2013 se crea en la Provincia de Mendoza un programa para la atención de los niños con asma, y el abordaje comunitario de esta patología. Entre las actividades educativas realizadas se dictaron pautas de manejo del asma. A pesar de ello existe una gran variabilidad en el manejo de la crisis asmática. Objetivo: Describir el manejo de las crisis de asma en niños en la provincia de mendoza, conocer la variabilidad entre los diferentes profesionales que atienden niños en atención primaria y guardias. Material y métodos: Se utilizó una encuesta anónima sobre supuestos casos clínicos de pacientes pediátricos con crisis asmáticas, distribuidos a médicos que atienden niños en nuestra provincia a través de los coordinadores regionales del Programa Provincial de Asma Infantil. Resultados: Se analizaron 112 encuestas. 53 médicos atienden en guardia polivalente, 35 en guardia de hospital pediátrico, 23 en centros de atención primaria. En la valoración de la gravedad de la crisis asmática en niños existe una alta coincidencia entre profesionales de los distintos niveles de atención. El uso de B2 con IDMp es la medicación indicada por más del 90% de los profesionales en la crisis independientemente de su gravedad como inicio de tratamiento de rescate. Menos del 30% utiliza corticoídes en las crisis leves y más del 80% utiliza corticoídes en las crisis moderadas y graves. No hay consenso en la utilización de una herramienta para valorar la gravedad de la crisis asmática. Conclusiones: Si bien existe concordancia en la valoración de la gravedad de la crisis asmática en niños solo el 36% de los profesionales utilizan escalas de gravedad y estos no tienen claro que escala utiliza. Este hecho refleja que a pesar de haber recibido capacitación y contar con una guía de manejo de asma, la instrumentación y utilización de la misma aún no es aplicada en forma adecuada en nuestro ámbito de trabajo.

151 IMPACT OF ROUTINE SPIROMETRY IN THE FREQUENCY OF ACUTE PULMONARY EXACERBATION DIAGNOSIS IN CYSTIC FIBROSIS.

Carolina Silva Barboza de Aquino Mota; Luiz Vicente Ribeiro F. da Silva Filho; Joaquim Carlos Rodrigues; Pediatric Pulmonology Unit, Instituto da Criança, Hospital das Clínicas da Faculdade de Medicina da USP.

Objective of the study: To evaluate the impact of routine spirometry (every patient encounter) in the frequency of acute pulmonary exacerbation (APE) diagnosis in CF patients. Methods: All CF patients attending our outpatient clinic and able to perform spirometry were included in the study, after patient/caregiver consent. This prospective study compared results from the calendar year 2014 with the two previous years (2012–13). During the study period, patients underwent spirometry before each consultation, using a Koko Spirometer (Nspire Health/USA). A standardized protocol of clinical parameters and treatment in use was filled in all visits. The attending physician, not involved directly in the research, did the diagnosis of acute pulmonary exacerbation (APE) by using clinical and spirometric parameters. The frequency of APEs was defined as the number of APEs divided by the number of patient encounters. Data from the previous two years (when spirometric testing was performed at 6 months intervals) were obtained from medical records and APEs defined as the need for antibiotic treatment due to clinical deterioration. The Institutional Ethics Committee approved the study. Comparison of frequencies (visits, APEs) between the two periods was performed by paired T-test or Wilcoxon signed rank test. Results: Eighty patients were included in the study, mean age (SD) of 12.13 (3.43) years, 61.3% males. In 2014 they had a mean (SD) of 5.22 (1.92) visits/year, while in the previous two years they had 4.64 (1.63) visits/year, p = 0.02. The median of frequency of acute pulmonary exacerbations was 25% of the consultations in 2014, while it was 13.2% in the years 2012–13, p = 0.004. Among 117 APEs diagnosed in 2014, attending MDs declared that spirometry helped in the diagnosis in 106 (90.6%) of the occasions. Conclusions: Routine spirometry at each patient encounter increases the frequency of APE diagnosis in CF patients, with significant clinical implications.

152 EVALUATION OF CHLORIDE AND SODIUM IN SWEAT TEST AND THE CORRELATION WITH AGE IN CLINICAL SAMPLES FROM A REFERENCE CENTER

Alethée Gaimaraes Faria Gaimaraes Faria; Fernando Augusto de Lima Marson; Antonio Fernando Ribeiro; Carla Cristina Souza Goméz; Maria de Fátima Servidoni; José Duece Ribeiro; Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas.

Objective: To evaluate how the chloride varies with subjects’ age and if additional information can be achieved by measuring of the sodium in sweat test in a reference center. Material: There are analyzed 5627 exams realized in our reference center. Methodology: Conducted a retrospective survey of 5627 individuals to analyze the Sweat Test. Subjects had a mean age of 12.09 ± 17.82 (from 0 to 85.58 years). Results: There was correlation with sodium and chloride in the studied population (R2 = 0.8870, p < 0.0001) and distributed in age groups: 0 to 6 months (Group 1 — 645 individuals) (R2 = 0.8977); between 6 months and 18 years (Group 2 — 3855 individuals) (R2 = 0.8686); and greater than 18 years (Group 3 — 1066 subjects) (R2 = 0.8693). There were minor amounts of sodium (34.44 ± 20.59 vs. 37.69 ± 21.26) (p < 0.001), chloride (29.90 ± 25.71 vs. 33.32 ± 26.51) (p < 0.001) and Cl/Na (0.80 ± 0.23 vs 0.82 ± 0.25) (p = 0.004) in the male group (2997 subjects) than in females (2630), the opposite was observed for the weight sweat (0.17 ± 0.05 vs 0.16 ± 0.05) (p = 0.017). The sodium value was different for the individuals groups distributed according to the amount of chloride (namely, 0 to 30 mEq/L; 30 to 40 mEq/L; 40 to 60 mEq/L; > 60 mEq/L) (p < 0.001), being all groups different each other. Chloride has a low correlation with age of subjects (R2 = 0.1010, y = 25.5331 + 0.4625x) (p < 0.001), considering the groups divided by CF diagnosis; the correlation was maintained for sodium (R2 = 0.1240; y = 30.6953 + 0.4005x) (p < 0.001) and Cl/Na (R2 = 0.009; y = 5.1421-0.05225x) (p < 0.001). Conclusion: In the studied population, chloride and sodium correlate with age, and the sodium has stronger correlation with the chloride.

153 CLINICAL AND EPIDEMIOLOGICAL ASPECTS OF PATIENTS WITH CYSTIC FIBROSIS IN THE STATE OF PARÁ, BRAZIL

Valória de Carvalho Martins (Hospital Universitário João de Barros Barreto – Universidade Federal do Pará); Andrea Kelly Cristina Ribeiro dos Santos (Universidade Federal do Pará); Ida Vanessa Doederlein Schwartz (Universidade Federal do Rio grande do Sul).

Cystic fibrosis is the most common autosomal recessive exocrinopathy in Caucasian populations, with clinical manifestations including sinusitis, chronic airway infections, pancreatic insufficiency, abnormally elevated electrolyte levels in sweat, and obstructive azoospernia. Diagnosis is based on the presence of two pathogenic mutations in the CFTR gene and functional tests that confirm abnormal chloride transport in the affected
organs. The objective of this study was to report the clinical profile of 135 patients with cystic fibrosis from the Northern region of Brazil, emphasizing potential ethnic, geographic, socioeconomic, and healthcare-related influences on phenotype expression. Aims: The present study sought to investigate the clinical profile of CF patients in a population characterized by a high degree of miscegenation, with a particular focus on analyzing the diagnostic and clinical context of these patients. Methods: This was an observational, outpatient study using a convenience sampling strategy. An epidemiologic questionnaire was completed by means of a chart review and clinical assessment, always conducted by the same investigator. Results: Of the subjects included in the sample, 85.2% were brown and 80% had a late diagnosis, despite typical manifestations of the disease. Other characteristics of the sample included low household income, malnutrition, a low frequency of the p.F508del mutation (21.4%), and a high frequency of the p.M470V polymorphism (76%). Conclusion: Knowledge of the clinical variability of patients with cystic fibrosis, taking into account genetic and environmental factors, may facilitate the introduction of therapies and procedures better adapted to this population.

154 PERSISTENT PNEUMONIA: WHAT IS THE DIAGNOSIS?
Debora Tolaini Pinto Pimentel; Caren Ishikawa; Sonia Mayumi; Beatriz Neuhaus Barbisan; Clovis Eduardo Tadeu Gomes; Luiz Hirotoshi Ota
Universidade Federal de São Paulo (UNIFESP)
Objective: Report a case of persistent radiological opacity and respiratory symptoms in an adolescent. Case Report: An eleven-year-old previously healthy boy presented a one-year history of productive cough and episodic hyperventilation in a child with hypoventilation and hypersomnolence. Methods: Objective: Evaluate the treatment with non-invasive positive pressure ventilation in a child with hypoventilation and hyperpersomnolence. Results: The patient showed improvement in daytime somnolence (mESS < 9), with alveolar hypoventilation and suspicion of Rapid-Onset Obesity with Hypoventilation. Hypothamic Autonomic Dysregulation and Neural Tumor Syndrome (ROHHADNET), we evaluate the pre- and post-modified Epworth Sleepiness Scale (mESS) and treatment adherence. Results: A nine-year-old girl had a paravertebral thoracic ganglioneuroblastoma resection in 2014. Since the age of 4 years, she had a history of weight gain. In the last year, she gained 10 kg (BMI z-score 4.56), had no height gain, and had two hospital admission due to acute asthma exacerbation and hypernatremia. She develop daytime excessive somnolence (mESS = 16) without history of sleep deprivation or inadequate sleep habits. The investigation showed negative PHOX2B, hypernatremia (189 mmol/L), hypothyroidism, daytime hypercapnia (PaCO2 = 48 mmHg), and 37% of total sleep time with SpO2 < 90%. Non-invasive ventilation was initiated at RR 12 bpm, IPAP 15 and EPAP 6 cm H2O. After one month of therapy, the child showed improvement in daytime somnolence (mESS = 7). Investigations of other endocrine and autonomic abnormalities are under investigation. Discussion: The ROHHADNET syndrome is characterized by hyperfagia, rapid weight gain, hypothalamic abnormalities, such as hypothyroidism and inappropriate secretion of anti-diuretic hormone that can appear a few years after the initial symptom, autonomic dysregulation and associated neurologic tumor. Eventually, central hypoventilation develop in all patients requiring mechanical ventilation. In this child, we observed a complete normalization of daytime somnolence with non-invasive ventilation.

155 SUSPICION OF RAPID-ONSET OBESITY WITH HYPOVENTILATION, HYPOTHALAMIC, AUTONOMIC DYSREGULATION AND NEURAL TUMOR SYNDROME
Raquel Mascarenhas Freitas; Thiara Maria Gomes da Silva; Beatriz Neuhaus Barbisan; Clovis Eduardo Tadeu Gomes; Sergio Tufik; Gustavo Antonio Moreira; Universidade Federal de São Paulo
Objective: Evaluate the treatment with non-invasive positive pressure ventilation in a child with hypoventilation and hyperpersomnolence. Methods: We performed a chart review from a child followed at the Pediatric Pulmonology Division of Universidade Federal de São Paulo. In a patient with alveolar hypoventilation and suspicion of Rapid-Onset Obesity with Hypoventilation, Hypothamic Autonomic Dysregulation and Neural Tumor Syndrome (ROHHADNET), we evaluate the pre- and post-modified Epworth Sleepiness Scale (mESS) and treatment adherence. Results: A nine-year-old girl had a paravertebral thoracic ganglioneuroblastoma resection in 2014. Since the age of 4 years, she had a history of weight gain. In the last year, she gained 10 kg (BMI z-score 4.56), had no height gain, and had two hospital admission due to acute asthma exacerbation and hypernatremia. She develop daytime excessive somnolence (mESS = 16) without history of sleep deprivation or inadequate sleep habits. The investigation showed negative PHOX2B, hypernatremia (189 mmol/L), hypothyroidism, daytime hypercapnia (PaCO2 = 48 mmHg), and 37% of total sleep time with SpO2 < 90%. Non-invasive ventilation was initiated at RR 12 bpm, IPAP 15 and EPAP 6 cm H2O. After one month of therapy, the child showed improvement in daytime somnolence (mESS = 7). Investigations of other endocrine and autonomic abnormalities are under investigation. Discussion: The ROHHADNET syndrome is characterized by hyperfagia, rapid weight gain, hypothalamic abnormalities, such as hypothyroidism and inappropriate secretion of anti-diuretic hormone that can appear a few years after the initial symptom, autonomic dysregulation and associated neurologic tumor. Eventually, central hypoventilation develop in all patients requiring mechanical ventilation. In this child, we observed a complete normalization of daytime somnolence with non-invasive ventilation.

156 DIFFUSE PULMONARY LYMPHANGIOMATOSIS. CASE REPORT
Juan Carlos Torres Salas; Valera Carlos Moreno; Mariela Zanudo Aquire; Hector Nunez Pancar; Instituto Nacional de Salud del Niño (INSN).
Objective: To show our experience in the treatment of diffuse pulmonary lymphangiomatosis (DPL). Case report: The case reports of two children aged 6 years old that developed DPL are described. The first child had emphysema and both had recurrent cough, dyspnea and chylothorax. In both cases the diagnosis was made through multislice spiral tomography (MET) that describes a characteristic pattern and confirmed by lung biopsy. During his early follow they received diet rich in medium chain triglycerides but persisted with high expenditure of pleural drainage. Treatment with methylprednisolone pulses to the arrest of hemoptysis and chylothorax regression of achieving the removal of chest tube. Then propanolol and Bevacizumab were administrated. Discussion: DPL is a rare pulmonary lymphatic system disorder characterized by diffuse mediastinal soft tissue infiltration, pulmonary interstitial parenchymal infiltration, and pleuropulmonary effusion. Although pathologically benign, it is a progressive and fatal disease. The disease is believed to be congenital and the majority of cases are diagnosed in childhood. DPL is difficult to diagnose due to its presentation with nonspecific symptoms, and is misdiagnosed as asthma or other respiratory diseases. Yet, such lymphatic diseases have some imaging characteristics which help in the diagnosis, can best be detected by MET. There are no standardized treatment protocols or guidelines for affected individuals, various treatments have been reported as part of small series of patients. Recently, have reported encouraging results with drugs that inhibit, the production of vascular endothelial growth factor (VEGF), specifically the drugs propanolol, and bevacizumab. Systemic corticosteroids play an important role in the inhibition of angiogenesis, and these cases demonstrate that their use in pulses is useful in reducing the pleural spending at the start of treatment of DPL to enable the removal of chest tube.

157 THE ASSESSMENT OF THE USE OF ELASTIC THRACOABDOMINAL BINDER IN THE PRUNE BELLY SYNDROME – REPORT OF 3 CASES
Pediatric Pulmonology
FOCROZU

Objective: Assessment of the thoracoabdominal dynamic in children diagnosed with Prune Belly syndrome. Case report: Three infants were examined without sedation, awake and with continuous monitoring of physiological signals. Data were collected through the respiratory inductive plethysmography through Respitrace® equipment. The analog signals were converted to digital signals and the mean of respiratory rate of 60breaths per minute were analyzed before and after placing the elastic thoracoabdominal binder. Before placing the binder, the 3 infants showed phase angle of 170, 80 and 120 degrees, respectively. It is the highest degree, related to higher thoracoabdominal distortion. After placing the elastic thoracoabdominal binder, the phase angles measured were lower, however, for only the first infant, who had high values of thoracoabdominal asynchrony, significantly reduced the phase angle after the placement of the binder.

Discussion: Knowing the specific characteristics, there are better strategies regarding the treatment and conduct, positively contributing to the improved breathing efficiency and, consequently, to the impact on quality of life."

158 FOLLOW UP CLINICAL DATA FROM CYSTIC FIBROSIS PATIENTS DIAGNOSED BY NEWBORN SCREENING. Renata Rodrigues Guiran; Elizete Aparecida Lomazi; Antônio Fernando Ribeiro; Roberto José Negrão Neogueira; José Dirceu Ribeiro; Aline Cristina Gonçalves; Universidade Estadual de Campinas

Aim: Evaluating the nutritional status, respiratory and pancreatic involvement in children with Cystic Fibrosis (CF) from the National Neonatal Screening Program, when followed a Brazilian reference center. Material and Methods: Retrospective cohort study set up in a tertiary out patient reference center. Medical files of all 40 children in follow up (aged from 3 to 60 months old) were reviewed in October, 2015, and data from the last three visits were recorded. Nutritional status was assessed by measuring weight, height and body mass index (BMI). Pulmonary disease was recognized by occurrence of respiratory symptoms. Pancreatic insufficiency was detected by acid steatocrit and/or Fecalelastase 1 dosage. The first isolation of Pseudomonas aeruginosa and Staphylococcus aureus in or opharyngeal swab were recorded. For studying purposes children were classified in 5 groups according to age. Results: CF diagnosis median age was 91 days (range 66–56 days). Median follow-up period was 29 months (range 6.6 – 53.2 months). The Z score median values for weight/age, height/age and BMI/age were lower in the diagnosis period than during the latter follow up in all groups. The first identification of Staphylococcus aureus was at 8.23 months (range 3.4–14 months) and Pseudomonas aeruginosa was at 15.2 months (range 6.1–34 months). Pancreatic insufficiency diagnosis median age was 3.7 months (range 2.6–4 months) and identified in 95% of patients. Conclusions: The nutritional status, as measured by Z score of weight, height and BMI has improved along with the follow-up. Anthropometric values showed detriment around the time of first respiratory symptoms. All the patients presented respiratory symptoms during the follow up period. Pancreatic insufficiency was identified in all patients with the diagnosis being before the first three months of age in most patients.

159 MICROORGANISMS ISOLATED IN SWAB OROPHARYNGEAL OF THE PATIENTS WITH CYSTIC FIBROSIS DIAGNOSED AFTER NEWBORN SCREENING Aline Cristina Gonçalves (Graduate Program in Child and Adolescent Health, Department of Pediatrics, School of Medical Science); Renan M. Mauch (Graduate Program in Child and Adolescent Health, Department of Pediatrics, School of Medical Science); Elizete Ap Lomazi (Department of Pediatrics, School of Medical Sciences of Unicamp and Pediatric Gastroenterology Lab); José Dirceu Ribeiro (Department of Pediatrics, School of Medical Sciences of Unicamp and Pulmonary Physiology Lab); Carlos Emilio Levy (Department of Clinical Pathology, School of Medical Sciences of Unicamp and Division Clinical Pathol); Antonio Fernando Ribeiro (Department of Pediatrics, School of Medical Sciences of Unicamp and Pediatric Gastroenterology Lab).

Objective of the Work: To evaluate the frequency of isolation of microorganisms in patients oropharyngeal swab with cystic fibrosis (CF) attended at specific outpatient at Hospital de Clínicas da Unicamp from February 2010 to November 2015. Materials and Methods: Retrospective study of all appraisal microbiological tests carried out in microbiology sector of the Department of Clinical Pathology at the Hospital de Clínicas da Unicamp of patients with screening neonatal positive for CF, from the first appointment at the Hospital de Clínicas da Unicamp, in the last 5 years. Results: The study included 55 patients [F508del? (n = 23); F508del/ F508del (n = 07); F508del/G542X (n = 01); G542X/C7 (n = 01); patients with mutations non identified (n = 23)], and the average age at diagnosis 4.38 months (SD = 7.48 months; median = 2.66 months). Was performed the total of 891 routine bacterial culture diagnostic, with an average of 3.2 tests per patient / year. The age of the first bacterium isolation: mean = 5.87 months; median = 4 months; SD = 7.86 months) and S. aureus bacteria most frequently isolated (n = 31/55) in this population. 7 patients had 03 or more positive cultures for S. aureus/year and 3 patients for P. aeruginosa non mucoid / year. Conclusion: The isolation of S. aureus and P. aeruginosa non mucoid in patients derived from the Screening Neonatal was premature but with low index colonization.

160 OSTEOCHONDROMA IN ADOLESCENT: RARE MANIFESTATION IN COSTAL ARCHES. Marta Cristina Duarte (Universidade Federal de Juiz de Fora); Teresa Cristina Esteves (Universidade Federal de Juiz de Fora); Teresa Cristina Ribeiro Lopes (Universidade Federal de Juiz de Fora); João Paulo Vieira (Hospital Regional João Penido); Bryan da Silva Marques Cajujo (Universidade Federal de Juiz de Fora); Ana Luiza Souza Dutra (Universidade Federal de Juiz de Fora);

Aim: To report the case of an adolescent with a rare thoracic tumour. Case Report: LHRS, 14 years old, female, white, presented with productive cough being before the first three months of age in most patients.
deformity. Surgical resection is indicated when there are symptoms or in order to reach a diagnosis. The importance of knowledge of thoracic bone tumors in the differential diagnosis of lung diseases must be considered.

161 MICROORGANISMS ISOLATED IN NEWBORNS IN THE SPECIALIZED HOSPITAL ASSISTANCE

Gabriela Taveira Estrela (UNIFAN); Thais Haddad Silveira (UNIFAN); Fabrício Ribeiro de Campos (Santa Casa de Franca – SP); Maria Auxiliadora Mancilha Carvalho Pedegone (UNIFAN); Marisa Afonso Andreu Brunhoroti (UNIFAN); Regina Helena Pires (UNIFAN)

Goal: Identify the microorganisms isolated in the sectors providing assistance to newborns, as well as the antimicrobial sensitivity profile. Method: Descriptive study held in 2014 in tertiary-level public hospital. Microorganisms identified from blood of newborns accompanied in Intensive Pediatric Therapy Center were considered, as well as internal and external nursery, being excluded the results of children from areas of Pediatrics and rooming. It was also examined the sensitivity of bacteria identified to local use antibiotics. Data were expressed as a percentage. Result: Sixty-two microorganisms have been identified. Among newborns in intensive care Center assistance, we have identified: Klebsiella pneumoniae (6%), Pseudomonas aeruginosa (10%), Enterobacter sakazakii (2%), Klebsiella ozaenae (2%), Staphylococcus coagulase (8%), Staphylococcus hominis (4%), Acinetobacter lwoffi (2%), Methicillin-resistant Staphylococcus aureus (MRSA) (38%) and Candida albicans (28%). In the nursery, external K. pneumoniae (50%) and Staphylococcus MRSA (50%) were identified, and the internal nursery the presence of Escherichia coli (33.3%), K. pneumoniae (33.3%) and Staphylococcus MRSA (33.3%) was found. Analyzing the bacterial sensitivity profile, it was found that S. aureus MRSA was the microorganism of greater strength, followed by K. ozaenae and E. sakazakii. Conclusion: Identified microorganisms show an antimicrobial resistance which may cause risk to the group of newborns. In this way, preventive actions may be directed in the pursuit of protection of child health. The adoption of protocols of rational use of antimicrobials is essential to avoid induction of bacterial resistance.

162 CASE REPORT: PNEUMOMEDIASTINUM AS A COMPLICATION IN PATIENTS WITH UNCONTROLLED ASTHMA

Eduardo Augusto Caldeire Storti; Maria Luiza da Costa Bertolini; Sandra Lange Zaponi Melek; Caroline Cecy Kuenzer Caron Fukashima; Hospital Angelina Caron.

Objective: The objective is to present a case report of pneumomediastinum and subcutaneous emphysema in a patient with asthma untreated. Case report: Patient VAM 7 year-old male was admitted to the emergency room complaining of shortness of breath, wheezing and fever for one day. On physical examination the child was in good general health, lucid and oriented, presence of edema in face respirator with wheezing, diffuse rales and oropharynx with abundant subsequent dripping. The palpation of the right thorax and cervical area showed subcutaneous cracking, consistent with subcutaneous emphysema findings. His father had a history of asthma, rhinitis carrier mother. Reported frequent use of oral corticosteroids in crises, but without monitoring with pediatric pulmonologist. In radiography and computed tomography was proven presence of pneumomediastinum and subcutaneous emphysema. The treatment was bronchospasm and sinus disease crisis, with good adherence and clinical outcome and decreased emphysema. He was hospitalized for 5 days return being oriented in one week, in which there demonstrating emphysema in clinical examination. Discussion: The report showed two unusual complications in patients with asthma: spontaneous pneumomediastinum and subcutaneous emphysema. The signs and symptoms vary due to the amount of air spaces found in mediastinal and severity of the disease. The diagnosis can be made only with chest X-ray, but with suspected differential diagnoses should be performed computerized tomography for further clarification. It is not advisable perform invasive procedures and tests in these cases only if patients have complications of more severely with the evolution to tension pneumothorax. The therapy used is conservative with rest and analgesia is required. Should investigate the triggering factor or cause for instituting adequate pre-treatment.

163 BRONCHOSCOPY AND BRONCHOALVEOLAR LAVAGE ROLE IN AN ANDEAN PEDIATRIC HOSPITAL: INITIAL EXPERIENCE

Angue-Martinez; Maria Eunestia; Villalba-Valencia Ximena; Zamarraga-Falcón Sonia; Jativa-Cruz Adriana; Enriquez- Marín Carolina; Zambrano-Palma Sandra; Hospital Pediatrico Baca Orti

Objective: we describe our experience in flexible and rigid bronchoscopy and its usefulness in bronchoalveolar lavage in children. Materials and methods: descriptive, retrospective and cohort study. Period: February/2014 to November/2015. We use rigid bronchoscopes from 3.0 x 26 to 5.0 x 30 and flexible videobronchoscope 5.4 with working channel of 2.2. Results: we realize 115 procedures. Age (years): median 10 (range 0.08–17), weight (kg): median 33 (range 3–53). Rigid bronchoscopies: 38 (33%) and flexible bronchoscopies: 77 (67%). Immunocompromised patients: 20 (17.9%) and immunocompetent patients: 95 (82.6%). Bronchoalveolar lavage by rigid and flexible technique: 105 (91%) with rescue bacterial and/ or fungal: 79 patients (75.2%). Main bacteria: Pseudomonas Aeruginosa (25.71%), Serratia Marcescens (12.3%), Klebsiella Pneumoniae (9.5%), Acinetobacter Baumanni (6.6%) and rescue mycological 9 cases (8.57%) all the samples were representative with presence of alveolar macrophages. The isolation of germ determined a change of medical conduct at 94.9% of cases.the main diagnoses that motivated the bronchoscopy were: nosocomial pneumonia: 39 (33.9%), bronchiectasis: 19 (16.5%) of which 4 patients with cystic fibrosis, persistent atelectasis: 18 (15.6%) and foreign bodies: 12 (10.4%), Procedence of patients: clinical areas: 33%, critical care unit: 22.7%, infectology room: 16.5% and emergency room: 9.5%. Complication major: 1.7%. Mortality: 0%.conclusions: both rigid bronchoscopy as flexible is useful in children and it serves as therapeutic and diagnostic tool. It is useful for the diagnosis etiologic of nosocomial infections. The rescue of microorganism is high in bronchoalveolar lavage and determined a change of conduct medical with good results.

164 CASE REPORT: LARYNGOTRACHEAL PAPILLOMATOSIS IN A INFANT

Milka Eugenia Monsalves Nilo (Escola de Saúde Pública do Ceará); Bernardo Paiva Júnior (Hospital Infantil Albert Sabio) Leopoldo Jacomel (SMS Fortaleza).

Objective: The purpose of this study is to present the case of a infant baby with recurrent laryngotracheal papillomatosis. Case report: Male infant, 5 months old, native from Pentecoste, Brazil, born by vaginal delivery at term, without complications. Family history of bronchial asthma (father) and genital condylomata (mother). When he was 3 months old baby, he began to suffer recurrent episodes of laryngeal stridor and respiratory discomfort, so he was sent to hospital of high complexity in Fortaleza, for a diagnostic investigation. A direct nasolaryngoscopy was done on him, which showed multiple papillomatous lesions in supra-glottis, in glottis, in atri-epiglottic ligament and in esophagus entrance. This patient was tracheostomized and several bronchoscopies were performed for multiple microsurgical resection. Furthermore, a histopathological study was done on the bronchoscopic material, which showed squamous papillomas with koylocotic changes. The patient evolved with frequent recurrences of stridor and respiratory distress, requiring hospitalization to perform serial bronchoscopy.

Pediatric Pulmonology
NEAR-FATAL ASTHMA IN ADOLESCENCE

Tânia Mara Baraky Bittar (Regional Hospital João Penido/FHEMIG, Juiz de Fora/Brazil); Marta Cristina Duarte (Meditação School of the Federal University of Juiz de Fora/Brazil); Teresa Cristina Ribeiro Lopes (Medical School of the Federal University of Juiz de Fora/Brazil); Ana Luíza Souza Dutra (Medical School of the Federal University of Juiz de Fora/Brazil); Sabrina Rodrigues Ribeiro (Regional Hospital João Penido/FHEMIG, Juiz de Fora/Brazil)

Aim: To report a case of near-fatal severe asthma in an adolescent, considering the risk factors. Case report: MERF, 13 years old, female, white, wheezing since she was an infant, which caused frequent visits to the emergency room and three hospitalizations for acute severe asthma. When she was five years old, she started taking high-dose beclomethasone and montelukast. However, it was difficult to control the symptoms. In August 2015, the prophylaxis was modified to budesonide / formoterol in individual capsules, having made irregular use of it. In parallel, emotional problems emerged and resulted in loss of asthma control, which made the patient start inhaled salbutamol, up to a bottle a week. In September 2015, she was admitted to the emergency room in respiratory arrest. After endotracheal intubation and stabilization, she was transferred to the pediatric ICU. During transportation she developed cardiopulmonary arrest, which responded to resuscitation maneuvers, and bilateral pneumothorax, properly detected and drained. She remained hospitalized for 40 days and was discharged with daily oral corticosteroids in addition to asthma prophylaxis. Discussion: Asthma is a controllable chronic inflammatory disease which may evolve to death if not treated properly. The risk factors in this case are numerous, such as adolescence, emotional changes, changing the inhalation device, poor treatment adherence, lack of skills to perform the prophylaxis and the crisis protocol, and poor awareness of the severity of the crisis. The use of LABA is associated with risk of intubation, hospitalization and death. In contrast, SABA, in high dosage, are related to heart arrhythmia. The association of both drugs may have contributed to the near-fatal outcome. Outpatient follow-up is essential, optimizing the medications and adherence to treatment, since a year after an episode of near-fatal asthma, 10% of asthmatics evolve to death.

BRONCHOSCOPY IN PEDIATRIC AND NEONATAL INTENSIVE CARE UNITS

Angelica Maria Barba Rueda (Programa de Pós-Graduação em Ciências Pneumológicas, Universidade Federal do Rio Grande do Sul – UFRS); Ana Cristina dos Santos Sabrito (Programa de Pós-Graduação em Ciências Pneumológicas, Universidade Federal do Rio Grande do Sul – UFRS); Julio Oliveira Espinel (Programa de Pós-Graduação em Ciências Pneumológicas, Universidade Federal do Rio Grande do Sul – UFRS); Cristiano Feijó Andrade (Universidade Federal do Rio Grande do Sul – UFRGS)

Introduction: The use of flexible bronchoscopy has been increased in critically ill patients with a wide range of therapeutic options such as removal of secretion in children with acute lobar atelectasis, lung hemorrhage, as well as for intubation in patients with difficult airways. Additionally, the obtaining of bronchoalveolar lavage (BAL) during bronchoscopy can help in the diagnosis of ventilator-associated pneumonia and lung infections in severe patients. Objectives: To evaluate the main indications of bronchoscopy and the yield of the bronchoalveolar lavage in pediatric patients from the pediatric ICU and neonatal ICU of the Santa Casa de Porto Alegre Hospital Complex. Methods: The medical records of patients who underwent bronchoscopy between January 2005 to June 2015 from the pediatric and neonatal ICU of the Santa Casa de Porto Alegre Hospital Complex were analyzed. Records with incomplete data were excluded. Results: 285 procedures were performed in 169 patients: 135 (80%) patients were from the pediatric ICU, with an average weight of 8.132 kg. 162 patients had some comorbidity and 49% had heart disease with 65% of heart surgery in these patients. Flexible bronchoscopy was performed in 92% and rigid bronchoscopy in 8%. 146 (86%) bronchoscopies were for diagnosis and 12% were therapeutic and diagnostic. The main indications for bronchoscopy were extubation failure in 69 (41%) patients, stridor in 25% and bronchoalveolar lavage in 24%. The most common finding was laryngomalacia in 50 patients (30%) being the most frequent alteration in patients whose indications were difficult and extubation and stridor. BAL was performed in 56% of the patients, with no bacterial identification in 61 samples (64%). The complication rate was 3%, with the most frequent complication being transient hypoxemia (2%). Conclusion: Bronchoscopy is a procedure that can be safely performed in critically ill patients and has good diagnostic and therapeutic efficacy.

CASE REPORT – BRONCHIOLITIS ACUTE VIRAL WITH COMPLICATED BRONCHIOLITIS OBITERANS

Tuanni Vanessa Werle (Hospital Infantil Pequeno Anjo); Gastão Dias Junior (Hospital Infantil Pequeno Anjo); Mariana Zamprogo Tezza (Hospital Infantil Pequeno Anjo); Jahumer Frantchesa Antal dos Santos (Hospital Infantil Pequeno Anjo); Karina Heusser (Hospital Infantil Pequeno Anjo); Vinicius Oro Popp (Universidade do Vale do Itajaí)

E.G.M.S., 7 months, male, white, prenatal and neonatal period was uneventful, previously healthy. Brought to the emergency with cough and wheezing compatible with acute viral bronchiolitis. Hospitalized due to respiratory distress and recurring goings to the health unit. In the second week he developed acute respiratory failure and fever, requiring submission to the Intensive Care Unit. Due to the clinical picture of wheeze, respiratory distress and dependence on supplementary oxygen therapy, the possibility of bronchiolitis obliterans (BO) has been raised. Chest tomography was performed and revealed a pattern of mosaic perfusion, consistent with BO. Excluding the diagnosis of cystic fibrosis, the treatment was initiated with daily respiratory physiotherapy, acetylcysteine, Azithromycin and inhaled corticosteroid therapy with fluticasone. Due to little improvement of breathing, the montelukast was associated and pulsetherapy with methylprednisolone. Progressively the patient improved breathing pattern. After the second pulsetherapy, the patient no longer needed oxygen therapy continuously and had dyspnea on exertion only. Bronchiolitis obliterans is an inflammatory disease of the small airways that occurs due to a previous injury to the lower respiratory tract. The persistent inflammation and fibrosis in terminal and respiratory bronchioles lead to a partial or total obliteration of the air in these locations and, subsequently, to chronic airway obstruction. 1 This disease presents different etiologies, but in children the most prevalent cause is in post-infectious situations. The diagnosis of BO is basically clinical and radiological. Its clinical picture is based on tachypnea and crackles/wheeze on auscultation, with persistent symptoms. In chest CT shows characteristic the mosaic pattern of perfusion. 2 There is no universal consensus on the treatment of BO, but therapeutic measures generally aim to reduce local inflammation.
Discussion: Spontaneous pneumothorax (SP) is a rare complication with shown a successful treatment with no recurrence of the complaints. Surgical thoracic drainage with subsequent pleurodesis. Follow-up has bilateral subpleural blebs at the apex of both lungs. Patient underwent new the upper lobe of the left lung. A new CT scan demonstrated multiple spaces. No signs of venous engorgement. X-ray showed a pneumothorax of hemithorax, with a concomitant hypertympanic percussion of the intercostal surgical drainage. At the prior admission, no CT scan abnormalities were admission due to a spontaneous pneumothorax of the right lung, treated by inhaled corticosteroids, with satisfactory control. History of recent hospital symptoms such as fever or fatigue was observed. Asthmatic, in use of three days. Parents denied additional respiratory symptoms. No systemic in sequential CT scan. Case Report: K.A., female, 12yo, came in for an recurrent spontaneous pneumothorax associated with blebs formation found in patients without underlying or overt lung disease, except for small subpleural blebs (blebs), while the secondary complication arises as previously known lung disease. The incidence of primary spontaneous pneumothorax is 6–10 cases per 100,000 inhabitants per year. The possibility of relapse from the first episode is around 30% and after the second episode it is 60 to 80%, decreasing the latency period. The diagnosis is based on the history and physical examination and confirmed by carrying out imaging methods. The radiograph obtained during forced expiration can aid in the visualization of small volumes of air in the pleural space, which are not visible in conventional radiography. Computed tomography of the chest can be valuable in special clinical situations, such as loculated pneumothorax and patients in intensive care unit. 2 The treatment of pneumothorax is very variable and depends on numerous factors. Choosing the most appropriate option depends on the intensity of symptoms, clinical repercussion, etiology and associated comorbidities. In all cases the main objectives are: to release the air in the pleural space, restore lung function and decrease the probability of recurrence.

RECURRENT SPONTANEOUS PNEUMOTHORAX ASSOCIATED WITH BLEBS FORMATION IN AN ASTHMATIC PATIENT: A CASE REPORT
Eduardo Augusto Caldeira Storti; Rafaela Wagner; Paulo César Kussek; Lorena Xavier Costa Brzezinski; Fernanda Henriques Lima e Silva Gois; Carolina Cardoso de Mello Prando; Hospital Pequeno Príncipe.

Objective: Report a case of spontaneous pneumothorax. Case report: Female patient, 1 year 10 months age, with dyspnea associated with coughing, grunting and abdominal pain. Physical examination revealed decreasing of vesicular murmur on the left. A chest x-ray was performed and revealed pneumothorax in the left side. Then, the patient was submitted to thoracostomy with drainage in water seal. The patient presents good evolution of the clinical picture and the thoracic drain was removed on the 4th day after surgery. An x-ray control was performed and it revealed left pneumatocele. The pneumothorax is defined by the presence of air accumulation in the pleural cavity, ranked spontaneous (primary or secondary) and not spontaneous. Primary spontaneous pneumothorax occurs in patients without underlying or overt lung disease, except for small bubbles subpleural (blebs), while the secondary complication arises as previously known lung disease. The incidence of primary spontaneous pneumothorax is 6–10 cases per 100,000 inhabitants per year. The possibility of relapse from the first episode is around 30% and after the second episode it is 60 to 80%, decreasing the latency period. The diagnosis is based on the history and physical examination and confirmed by carrying out imaging methods. The radiograph obtained during forced expiration can aid in the visualization of small volumes of air in the pleural space, which are not visible in conventional radiography. Computed tomography of the chest can be valuable in special clinical situations, such as loculated pneumothorax and patients in intensive care unit. 2 The treatment of pneumothorax is very variable and depends on numerous factors. Choosing the most appropriate option depends on the intensity of symptoms, clinical repercussion, etiology and associated comorbidities. In all cases the main objectives are: to release the air in the pleural space, restore lung function and decrease the probability of recurrence.

RECURRENT SPONTANEOUS PNEUMOTHORAX ASSOCIATED WITH BLEBS FORMATION IN AN ASTHMATIC PATIENT: A CASE REPORT
Joanna Vanessa Werle (1); Gastão Dias Junior(1); Jahumer Franthesca Antal dos Santos (1); Karina Heusser (1); Mariana Zanpregno Tezza (1); Vinicius Oro Popp (Universidade do Vale do Itajaí). 1-Hospital Infantil Pequeno Anjo

Objective: To report a case of spontaneous pneumothorax. Case report: Female patient, 1 year 10 months age, with dyspnea associated with coughing, grunting and abdominal pain. Physical examination revealed decreasing of vesicular murmur on the left. A chest x-ray was performed and revealed pneumothorax in the left side. Then, the patient was submitted to thoracostomy with drainage in water seal. The patient presents good evolution of the clinical picture and the thoracic drain was removed on the 4th day after surgery. An x-ray control was performed and it revealed left pneumatocele. The pneumothorax is defined by the presence of air accumulation in the pleural cavity, ranked spontaneous (primary or secondary) and not spontaneous. Primary spontaneous pneumothorax occurs in patients without underlying or overt lung disease, except for small bubbles subpleural (blebs), while the secondary complication arises as previously known lung disease. The incidence of primary spontaneous pneumothorax is 6–10 cases per 100,000 inhabitants per year. The possibility of relapse from the first episode is around 30% and after the second episode it is 60 to 80%, decreasing the latency period. The diagnosis is based on the history and physical examination and confirmed by carrying out imaging methods. The radiograph obtained during forced expiration can aid in the visualization of small volumes of air in the pleural space, which are not visible in conventional radiography. Computed tomography of the chest can be valuable in special clinical situations, such as loculated pneumothorax and patients in intensive care unit. 2 The treatment of pneumothorax is very variable and depends on numerous factors. Choosing the most appropriate option depends on the intensity of symptoms, clinical repercussion, etiology and associated comorbidities. In all cases the main objectives are: to release the air in the pleural space, restore lung function and decrease the probability of recurrence.
mix model statistics was used, with LF indices as response variables and time as as predictor variable. The intercept and time were assessed as fixed effects. Results: Mean values of ten points assessed were: FVC, 68.8% ± 17.7%; FEV1, 48% ± 15; FEV1/FVC, 66% ± 17, and FEF25-75, 25.4% ± 14. At long-term, there was a clinical and statistically significant improvement in FVC (p = 0.04). Both FEV1 and FEF25-75, which best reflect the obstructive component, did not show significant changes clinically or statistically (p=7.08 and p = 0.873 respectively), maintaining similar values in percentage of predicted over time. FEV1/FVC showed a statistically significant change (p = 0.015), although clinically explained by greater improvement of FVC compared to FEV1. Conclusions: The results of our study suggest that, in children and adolescents with post-infectious bronchitis obliterans, the obstructive component of pulmonary function (FEV1 and FEF25-75) did not show significant changes over time, either deleterious or beneficial, over time. The forced vital capacity, on the other hand, showed a progressive increase over time that may be of great importance as a overall protection factor once the physiologic decline of LF starts in adulthood.

172 FOLLOW-UP OF SIX MINUTE WALK TEST IN CHILDREN AND ADOLESCENTS WITH CYSTIC FIBROSIS

Maira Bentes de Almeida Ramos; Lívia Cristina Avelino Costa; Bruna de Souza Sixel; Ana Lúcia Nunes Diniz; Christine Pereira Gonçalves; Nélbe Nesi Santana; Instituto Nacional de Saúde da Mulher, da Criança e do Adolescente Fernandes Figueira

Objective: The 6-minute walk test (6MWT) is a submaximal exercise test used to assess functional capacity, response to treatment and prognosis in cardiopulmonary diseases. This study aimed to compare the 6MWT performance after 12 months of outpatient follow-up in children and adolescents with cystic fibrosis. Methods: We analyzed 23 pairs of 6MWT performance after 12 months of outpatient follow-up in children and adolescents with confirmed diagnoses of Cystic Fibrosis from January 2012 to November 2015. All patients were followed by respiratory physiotherapy. The 6MWT were performed with an interval of 12 months, according ATS recommendations. Measured outcome was the distance walked. Continuous variables are presented as mean and standard deviation. T-paired test was used to compare means. Significance was defined as p-value <0.05. Results: In our sample, 9 were male. Age at the first test was 10.39 ± 3.99 years. After 12 months, the weight, height and peak expiratory flow rate increased. The observed values were, respectively, 33.76 ± 11.85 and 35.5 ± 12.03 Kg; 1.39 ± 0.20 and 1.43 ± 0.20 m; 218.7 ± 93.22 and 253.7 ± 103.28 L/min (p < 0.05). No significant differences were found for the other variables. The walked distance was 99.4 ± 12.5 and 98.5 ± 11.6% of predicted. The subjective perception of dyspnea and lower limbs fatigue were respectively 1.3 ± 1.4 and 0.9 ± 1.3; 1.8 ± 1.5 and 1.5 ± 1.5. Conclusion: The present study shows that there isst decline in 6MWT performance after 12 months of follow-up in children and adolescents with cystic fibrosis.

173 NECROTIZING PNEUMONIA – REPORT OF A CASE

Gabriela Sousa Moreira; Bruna Eulârio Castanheira; João Batista Salomão Junior; Stella Maria de Almeida e Silva; Douglas Lopes Vieira Arantes; Ana Paula da Silva Rodrigues; Hospital da Criança e Maternidade de São José do Rio Preto

Necrotizing pneumonia is a rare complication of lobar pneumonia, and is characterized by extensive necrosis and liquefaction with lung tissue cavitation. Although being severe in the acute phase, when the disease is properly diagnosed and well conducted, its prognosis is favorable in the pediatric age, according to the following case. Two year old patient, male, was admitted in the emergency service with fever and vomiting one week ago. One day before, the patient was in hypotensive state, with grunting and fatigue. At physical examination, patient presented a regular general state, prostrate, bleached, slightly dehydrated, afebrile. Vesicular murmur decreased in the right base with signs of respiratory effort. Chest radiograph with opacification of the hemithorax at right. Complementary exams presented unchanged blood count, but C-reactive protein of 29.39 mg/dL. A combined antibiotic therapy was started and thoracentesis was also performed. The patient evolved with respiratory failure and a orotracheal intubation, intensive care and chest drainage were required. Pleural fluid was positive for Streptococcus pneumoniae. Nine days later, the patient was submitted to exploratory thoracotomy with right upper and lower pulmonary lobectomy. After the procedure, the patient was maintained in intensive care, with vasoactive drugs and extended antibiotic therapy. A gradual evolution was evidenced, accompanied with improvement of the clinical state. S. pneumoniae is the predominant etiologic agent. When the child with pneumonia presents prolonged fever without any improvement associated with antibiotics or septicemia, the diagnosis probably should be revised. Also, diagnosis is confirmed by X-ray or CT scan of the chest. Pleural fluid is usually present. The clinical treatment should be instituted during a long period. Surgical interventions, such as pleural drainage and thoracotomy, may be requested in selected cases. Proper management is crucial for a favorable outcome.

174 BRONCHOGENIC CYST – CASE REPORT

Tuanvi Vanessa Werle (1); Gastão Dias Junior (1); Mariana Zampogno Tezza (1); Karina Heusser (1); Jahumer Frantchesca Antal dos Santos (1); Gilberto A. Tesser Augusto (Universidade do Vale do Itajaí), 1-Hospital Infantil Pequeno Anjo

Patient named Y.M.S.P., female, nine months and four days, native of Itajaí-SC, with a respiratory infections history since the first months of life. She achieved four hospitalizations and X-ray images showed only diffuse alveolar opacification in the right hemithorax. It was suspected also pulmonary malformations, so, a investigation by chest tomography was conducted. Subsequently, the MRI elucidated a cyst with probable origin in the middle mediastinum, extending into the left hemithorax. Its main diagnostic hypothesis was the possibility of bronchogenic cyst and esophageal duplication cyst. At 1 year and 2 months of life she was transferred to the Pediatric Surgery Department of JGCH, when the diagnosis of BC had been confirmed by surgeons. A posterolateral left thoracostomy was performed with cyssectomy measuring 3 cm in the largest diameter. Macroscopy showed cystic lesion mucoid compatible with BC. The patient recovered well after surgery and became asymptomatic. Review BC is the most common cystic lesion of the mediastinum. It presents estimated incidence of 6 to 15% of primary mediastinal masses.2 The BC originates from primitive intestinal structures and results from abnormal development of embryonic tracheo-bronchial tree. In infants, it is common respiratory failure, with compressive symptoms of mediastinal structures, while in adults and children, recurrent respiratory infections are prevalent. 4 The CB usually presents with antibiotics or septicemia, the diagnosis probably should be revised. 5Currently, surgical treatment with complete resection of the cyst by thoracotomy or video-assisted thoracoscopy is indicated for all symptomatic BC. Meanwhile, the surgery may be an option in asymptomatic cases, since 85% of these lesions may become symptomatic over time. 6

175 PEDIATRIC PULMONOLOGY CLINIC: EVALUATION OF 257 FIRST-ATTENDANCE RECORDS, WITH EMPHASIS ON ANAMNESIS AND CLINICAL EXAMINATION IMPORTANCE

Pediatric Pulmonology
Talitha Di Martha Chacon Belinati; Débora Tolaini Pinto Pimentel; Edson Taípina Braga; Carlos Roberto Bazzo; Clóvis Eduardo Tadeu Gomes; Universidade Federal de São Paulo- UNIFESP.

Goal: To correlate the diagnoses, their referrals and hypotheses assessed in the specialty clinic (third-party attendance), based exclusively on anamnesis and clinical examination, evaluating the detected pulmonary pathologies and their procedures. Material and methods: Descriptive observational study. The data was collected from 257 first-attendance standard records in the Universidade de São Paulo Pediatric Pulmonology Clinic, from 2007 to 2009. The following items were evaluated: gender, age, weight, height, atopy family history, as well as the diagnoses, their referrals and detected pathologies, together with their respective procedures. Results: Out of 257 patients, 97 children (37.7%) were referred with “wheezing”, “bronchoconstriction”. Out of these, 79 cases (81%) received asthma diagnosis after the first attendance. Out of the cases referred with recurrent pneumonia, 48% had the asthma diagnosis. Out of the obtained procedures, 20% of the total cases were discharged from the service after the specialist first evaluation, and 47% of the cases were treated with inhaled corticosteroids at first. In this assessment, it was also observed that 64% of the referred patients presented atopic mothers and 27% atopic fathers. Conclusion: Asthma is the most frequent infant pulmonary pathology and a more detailed anamnesis must be considered before referring the patient to a specialty, avoiding unnecessary and expensive referrals, once the asthma diagnosis depends more on the anamnesis and clinical examination and less on laboratory or radiologic examinations.

CHEST LYMPHANGIOMA
Renata Pimenta Buzatto; Paulo Cesar Kassek; Hospital Pequeno Príncipe.

Objective: Report case of a patient with recurrent pleural effusion due to thoracic lymphangioma. Case Report: A 2-year-old patient (female) with acute respiratory failure due to bilateral pleural effusion was hospitalized in pediatric hospital. The caregiver said that the patient had previously hospitalization by pneumonia and pleural effusion treated with antibiotics and pleurodesis. A workup revealed sweat test borderline in two samples, chest X-ray opacity compromising the lower third of the right hemithorax. Sputum culture was positive to Staphylococcus aureus. The liquid cavity analysis showed increased lactate dehydrogenase and a slight increase in a total protein. The tomography revealed expansive cystic lesion, septated in thoracoabdominal transition, with a thick content, encapsulated measuring about 14 × 9 × 8 cm. A surgical extraction was done but it was not possible to complete withdrawal of the mass because there was infiltration in the abdomen. The histological examination was consistent with lymphangioma. Today, the patient is asymptomatic, without relapses and without medication. Discussion: Cystic lymphangioma is one of several primary mediastinal lymph and lung disorders and is defined as a congenital malformation of the lymphatic system than a true neoplasia. It is a extremely rare benign tumor, slow growing and usually asymptomatic until it reaches large or be subject to malignant disease. These authors suggest that patients without those features don’t need invasive tests. In our series, all patients had unilateral lymph node enlargement, without involvement of anterior mediastinum, lymphopenia, and enlargement of cervical lymph nodes are more likely to malignant disease. These authors suggest that patients with involvement of anterior mediastinum, lymphopenia, and enlargement of cervical lymph nodes are more likely to malignant disease. These authors suggest that patients with involvement of anterior mediastinum, lymphopenia, and enlargement of cervical lymph nodes are more likely to malignant disease. These authors suggest that patients with involvement of anterior mediastinum, lymphopenia, and enlargement of cervical lymph nodes are more likely to malignant disease. These authors suggest that patients with involvement of anterior mediastinum, lymphopenia, and enlargement of cervical lymph nodes are more likely to malignant disease. These authors suggest that patients with involvement of anterior mediastinum, lymphopenia, and enlargement of cervical lymph nodes are more likely to malignant disease.

THYMIC HYPERPLASIA
Eugenio Fernandes de Magalhães; Marcus Vinícius Landin Stori Milan; Ana Paula de Oliveira Fernandes; Manuel Gouveia Otero Y Gomez; Isabela de Sousa Pereira; Anna Luiza Pires Vieira; Universidade do Vale do Sapucaí – UNIVAS

Objective: To report a case of thymic hyperplasia. Material and Methods: An infant with two months old, male, which started with anorexia, nasal congestion and hyaline rhinorrhea 5 days ago. In care, patient presented good general condition, no fever, and without signs of respiratory distress. For the other devices and systems changes were not disclosed. Chest radiography was performed, demonstrating radiopaque image in the right hemithorax. It was made a computed tomography scan of the chest showed a homogeneous mass, with well defined limits, in the region of the right anterior mediastinum, suggesting thymus hypertrophy. As conduct was suggested outpatient follow-up and guidance to the patient. Discussion: The thymus is a lymph organ located in the anterior mediastinum which plays a key role in the development and maturation of the immune system during childhood. It is a frequent source of confusion in the interpretation of chest X-ray especially in the differential diagnosis of mediastinal masses, cardiomegaly and pneumonia. Although the thymus growth continues during infancy to pre-pubertal, it is relatively more prominent in infants and young children. The classic radiological sign of thymus hypertrophy is the “sign of the candle “, as the thymus protrudes laterally to the upper mediastinum, and its remaining form resembles a ship sailing. The absence of air bronchograms help in the consolidation of distinction of the upper lobe. Conclusion: The thymic hyperplasia should be considered in the differential diagnosis of mediastinal masses and infectious conditions, mostly in pediatric population. Is necessary clinical and radiological follow-up of these patients.

178 MEDIASTINAL MASSES IN CHILDREN: A DIAGNOSTIC DILEMMA
Tania Wrobel Folescu; Renata Wrobel Folescu Cohen; Deborah Aragão Barroso de Pinho; Patricia Fernandes Barreto Machado Costa; Renato Farm D’Amoedo; Marjana Silva de Farias; Instituto Nacional de Saúde da Mulher, da Criança e do Adolescente Fernandes Figueira.

Objective: Describe diagnosis of pediatric patients referred for mediastinal mass investigation. Material and Methods: Review of the health records of 5 patients < 19years referred to the Pulmonology section from june to december 2015 for evaluation of mediastinal masses. Results: Mean age was 5.3 (0-13yo), 4/5 presented fever and 3/5 cough. Other symptoms/signs included malaise, weight loss and dyspnea (2/5) and bilateral wheezing (1/5). All patients had CT scan that showed unilateral solid mass, subsequently identified as lymph node enlargement (4/5) and broncoscopy revealed airway but no vascular compression (3/5). Histoplasma antibody was detected in 3 patients which were assumed as histoplasmosis and 2 of them were submitted to specific treatment. Tuberculosis was diagnosed and treated in 2 patients (1 with erythema nodosum;1 with heterogenous lymph node). Conclusion: Mediastinal masses in the pediatric population comprehends benign and malignant tumors, congenital anomalies and infections. Naeem et al described characteristics of mediastinal masses and concluded that patients with involvement of anterior mediastinum, lymphopenia, and enlargement of cervical lymph nodes are more likely to malignant disease. These authors suggest that patients without those features don’t need invasive tests. In our series, all patients had unilateral lymph node enlargement, without involvement of cervical lymph nodes and no lymphopenia. Brazil is one of the 22 countries that account for 80% of global tuberculosis cases and Rio de Janeiro has the highest incidence rate. Rio de Janeiro is responsible for most of the microepidemics of Histoplasma capsulatum infection described in Brazil. In our series, all patients came from endemic areas of tuberculosis and histoplasmosis and presented clinical-radiological criteria for both diseases. Distinguishing between benign and malign mediastinal masses is challenging in endemic areas, where half of the cases are attributable for these infections.

177 MULTILOCULATED PNEUMOMEDIASTINUM IN A NEWBORN: CASE REPORT
Pediatric Pulmonology
Background: To discuss a case of a newborn who presented multiloculated pneumomediastinum and its origin. Materials and Methods: This case report was based on medical record review, analysis of laboratory tests and imaging in addition to literature review. Results: We report a 41-week gestational age male newborn. The newborn showed ineffective ventilation from the first postnatal minute, and two cycles of positive pressure ventilation with a bag and mask was immediately started with good recovery. At 10 h after birth he developed respiratory distress (tachypnea, grunting and retractions). Physical examinations showed some diffuse crackles and hypophonetic heart sounds. Research to early neonatal infections and empirical antibiotic therapy were begun. Chest radiographs showed pneumomediastinum. A chest radiograph taken at 7 days old revealed a “spinnaker sail sign”. Chest computed tomography (CT) demonstrated multiloculated pneumomediastinum. Management was conservative. Discussion: Neonatal pneumomediastinum has been related to underlying lung disease or following atrumatic resuscitation at birth. Spontaneous pneumomediastinum in newborns is relatively rare. The evolution is usually benign and generally asymptomatic or oligosymptomatic with conservative treatment. Septation is a particularity of neonatal pneumomediastinum. The commonest diagnostic radiographic sign of pneumomediastinum is the “spinnaker sail sign”. When the chest X-ray is normal or doubtful, chest CT can be performed for better diagnosis. Conclusion: We conclude that the neonatal pneumomediastinum can be an under diagnosed condition as it manifests clinically as other common respiratory diseases. Spontaneous pneumomediastinum in newborns is a rare condition and must be sought secondary causes. Neonatal resuscitation is a possible etiology, but it is not possible to confirm this hypothesis since there were adequate support and qualified professional assistance.

### 180 IMPACT OF A PROLONGED VENTILATION UNIT IN PAEDIATRIC SERVICE

**Natalia Galas Souza (Universidade de Concepción); Daniel Zenteno Arros (Hospital Guillermo Grant Benavente); Camilo Barreza Etcheverry (Universidade de Concepción); Jaime Tapia Zapatero (Hospital Guillermo Grant Benavente); Ximena Andrea Paulette Navarro Tapia (Hospital Guillermo Grant Benavente).**

Objectives: The aim of this study is to measure the impact of a Paediatric Prolonged Ventilation Unit after three years functioning. Materials: 33 patients were included, transferred from the ICU between June 2012 and June 2015. Methodology: Descriptive statistic of general characteristics, time of hospitalization and conditions when discharged were analyzed. It was calculated the impact of a Paediatric Prolonged Ventilation Unit (PPVU) in critical bed occupancy, based on the change in the rotation index and occupancy in ICU, and the potential discharges that could have been made. Results: The entry age average was 4,6 years old (r = 0, 3–16); bpsys 18 (55%). Diagnosis: Neurological damage 43% (n=14), Neuromuscular disease 24 % (n=8), Chronic pulmonary disease 21 % (n=7), and Obstruction of superior airway track 12 % (n=4). total time of hospitalization 174 days (r = 5-1721) and in y en PPVU 57 (r = 2-783). There was no mortality. Most of the patients were discharged to their home 76% (n=25). Entered to ministerial programs 79% (n=26); Invasive ventilatory assistance 43% (14), Non invasive ventilatory assistance 50% (10). Oxygeen 58% (19). 55450 beds were occupied in the PPVU during the period; this allowed the entry of 125 children to the ICU. Te ocupational index in the ICU and the average of days of hospitalization was lower after the creation of the PPVU. Conclusion: The impact of the implementation of a PPVU is favorable; it increased the rotation of patients in the ICU and allowed the discharge of patients with complex respiratory disease.

### 181 WITHDRAWN.

### 182 VITAMIN D STATUS OF CHILDREN AND ADOLESCENTS WITH CISTIC FIBROSE

**Claudia de Castro e Silva (Hospital Infantil Albert Sabin/Universidade Federal do Ceará); Hildenia Baltazar Ribeiro Nogueira (Hospital Infantil Albert Sabin/Universidade de Fortaleza); Patrícia Sampio Padilha (1); Débora Pongitori Gifoni (1); Amanda Vale Catundal(1); Lia Almeida de Sá (1) L – Universidade De Fortaleza**

Objectives: To identify the vitamin D levels in patients followed at a specialized Cystic Fibrosis(CF); To associate vitamin D deficient values with the presence of pancreatic insufficiency and nutritional status. Materials/Methods: Retrospective, descriptive and quantitative study of the medical charts from 21 pediatric patients with cystic fibrosis accompanied at the Hospital Infantil Albert Sabin in Fortaleza–CE. The collection and data analysis took place between September and December 2015 by filling a standardized form. Sérum 25-hydroxyvitamin D(25-OHD) was measured with chemiluminescence enzyme immunoassay. The deficiency was defined as below 20 ng/ml, insufficiency of 20–30 and sufficiency of 30–50. For evaluation of pancreatic insufficiency use the test positivity SUDAMIII with higher reference values than 5% of fecal fat. Results: The 25-OHD status was measured in 21 patients. Of the patients analyzed, 47,6%(10) showed vitamin D insufficiency and 23.8%(5) deficiency of vitamin D totaling a percentage of 71.4%(15) of hypovitaminosis. Among them, it was identified growth rate reduced and low weight gain in 73.3%(11); 33.3%(8) were pancreatic insufficiency with changes in the analysis of SUDAM III and diarrhea; 80%(12) using a support dose of fat soluble vitamins. We identified 33.3%(5) of the sample with Body Mass Index <3 percentile; 66.6%(10) between 3 percentile and <85. Conclusions: A association was found between vitamin D deficiency and CF, reflecting on the low weight gain and growth deficit even in patients using fat-soluble vitamins in recommended doses. The genesis of these changes may be due to pancreatic insufficiency, sunlight deficiency or malnutrition. This shows the necessity of tracking the dosage of vitamin D and its adequate replacement. We stress the importance of adequate vitamin D dose in protocols for CF patients, as well as research and mentoring about the sun exposure of monitored patients.

### 183 CONGENITAL LUNG CYST MIMICKING ASTHMA

**Vitor Costa Palazzo (Hospital Infantil Pequeno Principe); Jürgen Beuther (1); Jéssica Takaki (1); Juliana Ardigó Lopes (1); Caroline Mika Shin-Ike (1); Andreza de Carvalho Formiga (1); I – Pontificia Universidade Católica do Paraná.**

Objective: To report a case of congenital lung cyst from a patient with symptoms of asthma. Case Report: Male patient, three months old, that had dyspnea on feedings, tachypnea and wheezing. No other changes in physical examination. Chest radiography showed rounded image with air content located in the right lower lobe. The hypothesis was acute viral bronchiolitis and lung cyst. He was treated with inhaled beclomethasone, montelucat, salbutamol to the crisis, and opted for outpatient follow-up of the lung cyst. With 1,5 years old, he was diagnosed with pneumonia and admitted to the emergency room with fever, productive cough and difficult for breathing. After partial improvement with antibiotic therapy, it was suggested the diagnosis of infected lung cyst and performed thoracotomy and right lower lobectomy – cyst of great size adhered to the pleura, with pus inside. The pathology confirmed an extensive necrotic area with abscessation. After removal of the cyst, the patient has been asymptomatic. DISCUSSION: Although rare frequency, the most common congenital malformation is bronchogenic cysts which may be of different sizes, single or multiple.
loculated or not. Common symptoms are dyspnea, cyanosis, cough, stridor, dysphagia, hemoptyis and chest pain. Most prevalent complications are intracavitary bleeding, recurrent pneumonia, bronchial obstruction, pneumothorax and tracheal compression. There are few data in the literature related to the presence of a cystic lesion with symptoms of bronchial asthma as the reported case. Symptoms similar to asthma result from partial compression of the trachea and bronchi and the occurrence of complete remission after cyst resection are present in all cases described, confirming the good evolution after surgery. The therapeutic method is the complete resection of the cyst, regardless of the presence of symptoms that establishes definitive histologic diagnosis, relieve symptoms and prevent complications.

184 NEONATAL CHOLESTASIS, SEVERE ANEMIA AND EDEMA AS MANIFESTATIONS OF CYSTIC FIBROSIS
Débora Carla Chong e Silva; Cristine Secco Rosário; Luís Keiko Lopes; Carlos Antônio Riedi; Nelson Augusto Rosário Filho; Universidade Federal do Paraná.
Objective: To report a case of cholestasis, severe anemia and edema as manifestations of Cystic Fibrosis (CF). Case Report: Two month-old girl from Prudentópolis, Southern Brazil, was admitted to investigate jaundice, anemia and manifestations of CF. She was referred to the Pediatric Pulmonology for follow-up. Discussion: PA is characterized by the presence of rudimentary bronchus measuring 5 mm. Newborn in good general condition, without complications, being discharged for outpatient treatment. Morphologic ultrasonography (USG) showing fetal dextrocardia. Pregnancy was打球 and was also precribed albendazole. With no improvement in the clinical condition of patient, new tests showed a total IgE of 5.4%. Possible mechanisms of such conditions include reduced pancreatic exocrine function, low dietary intake and reduced liver synthesis. Neonatal cholestasis in CF is a rare complication and meconium ileus is reported as a risk factor for its development. The outcome of CF patients presenting with neonatal cholestasis varies from full recovery within the first months of life in the majority to occasional cases of liver failure.

185 RIGHT PULMONARY APLASIA
Eugênio Fernandes de Magalhães; Amanda Vilela Breias; Amanda Pinto Botêga; Augusto Castelli Von Atzigen; Anna Luiza Pires Vieira; Universidade do Vale do Sapucaí – UNIVAS.
Objective: Relate a pulmonary aplasia case. Materials and methods: Pregnant woman born and resident in Pouso Alegre (MG) took a morphologic ultrasonography (USG) showing fetal dextrocardia. Pregnancy evolved to natural birth without complications. Female newborn, apgar of 9–10, asymptomatic. Physical examination: Heart sounds and stroke to the right and vesicular murmur on the right hemithorax. Scoliosis chest x-ray that found radiopacity to the right with cardiac silhouette deflected to the same side. It was thus elaborated the diagnosis of aplasia/pulmonary agenesis. Echocardiogram conducted at 2 days old revealing dextrocardia, situs solitus, patent ovale foramen and right aortic arch. Newborn remained asymptomatic. Traffic study also performed with contrasted x-ray that found radiopacity to the right with cardiac silhouette deflected to the right and vesicular murmur on the right hemithorax. Solicited chest x-ray showed intracavitary bleeding, recurrent pneumonia, bronchial obstruction, pneumothorax and tracheal compression. There are few data in the literature related to the presence of a cystic lesion with symptoms of bronchial asthma as the reported case. Symptoms similar to asthma result from partial compression of the trachea and bronchi and the occurrence of complete remission after cyst resection are present in all cases described, confirming the good evolution after surgery. The therapeutic method is the complete resection of the cyst, regardless of the presence of symptoms that establishes definitive histologic diagnosis, relieve symptoms and prevent complications.

186 CLINICAL-FUNCTIONAL PROFILE OF CHILDREN AND ADOLESCENTS WITH ASTHMA IN A SPECIALIZED CLINIC IN THE MIDWEST REGION OF BRAZIL
Warley Diogo Francisco Duarte; Marília da Silva Garrote; Rafaela Furguro Baragatti; Lusmaia Damaceno Camargo Costa; Raquel Vidica Fernandes; Isadora de Oliveira Cavalcante; Clinical Hospital of Federal University of Goiás (UFG).
To analyze clinical and functional profile of asthmatic children and adolescents in an outpatient treatment. Methods: Cross-sectional study with data collected from medical records of children and adolescents with asthma treated at Clinical Hospital of Federal University of Goiás (UFG), between Jan-Nov2015. Clinical data of the first and last visit were collected and those patients who had done a spirometry, anthropometric and clinical data were collected on the day of the exam. The study was approved by Medical Ethics Committee. Statistical analysis was performed using SPSS 23.0. Results: There were 254 patients in that period, 36 were excluded due to difficulties in recovering data records. The median age was 8 years (10.0–18.0), the most prevalent age group was 5–11 y (n = 123, 56.7%) where most patients were male (59.9%). The majority of patients (62.6%) had adequate nutritional status (Z-score –2 and +1), followed by overweight and obesity (18% and 10.9%, respectively). Regarding the severity levels, 45.4% of patients had moderate and 20.6% severe asthma. Most patients (79.4%) were taking medium/high doses of inhaled corticosteroids, 20.6% anti-leukotrienes and 27.5% long-acting bronchodilator for asthma control. However, only 17.4% were well controlled and 53.7% partly controlled. The main reason for not achieving the disease control was due to poor adherence (25.0%) and incorrect handling of the inhaler, occurring in 8.8% of cases. Most patients (94.4%) had allergic rhinitis (AR), 62.6% had persistent moderate to severe AR. On a functional evaluation (n = 106), VEF1 and VEF1/CVF were normal in 61.3% and 68.9%, respectively. Conclusions: Most patients had moderate to severe asthma taking medium/high doses of inhaled corticosteroids and had unsatisfactory clinical control despite adequate functional parameters. Unsatisfactory adherence to treatment was the main reason for suboptimal control of the disease.

187 LOEFFLER SYNDROME IN THE DIFFERENTIAL DIAGNOSIS OF SEVERE ASTHMA
Débora Carla Chong e Silva; Yuri Rosado Care de Medeiros; Elio Issao Kametani; Ronaldo Gustavo Albini Tyski; Wagner Granelli Junior; Maria Fernanda Gasparoto Tonin; Pontifícia Universidade Católica do Paraná.
Objective: To report a case of a patient with Loeffler syndrome, who has been infected by the Toxocara canis parasite and the importance of remembering this disease as differential diagnosed with severe asthma. Case Report: Male, 7 years and 11 months, with difficult controlling asthma. Patient was using inhaled budesonide and formoterol and aminophylline and associated with renal and airway malformation. The clinical manifestations range from asymptomatic to severe respiratory failure after birth. Asymptomatic patients do not require intervention, however, they must prevent themselves from pulmonary infections and treat them beforehand, improving the diagnosis and their quality of life. Prenatal diagnosis helps prevent birth complications and a bigger neonatal support. Thus, advances in the early diagnosis and interventions help to achieve a better prognosis. Unlike literature, in this case there was no association with urinary tract and respiratory malfunctions, and PA went to the right side. It was thus elaborated the diagnosis of aplasia/pulmonary agenesis. Echocardiogram conducted at 2 days old revealing dextrocardia, situs solitus, patent ovale foramen and right aortic arch. Newborn remained asymptomatic. Traffic study also performed with contrasted x-ray that found radiopacity to the right with cardiac silhouette deflected to the right and vesicular murmur on the right hemithorax. Solicited chest x-ray showed intracavitary bleeding, recurrent pneumonia, bronchial obstruction, pneumothorax and tracheal compression. There are few data in the literature related to the presence of a cystic lesion with symptoms of bronchial asthma as the reported case. Symptoms similar to asthma result from partial compression of the trachea and bronchi and the occurrence of complete remission after cyst resection are present in all cases described, confirming the good evolution after surgery. The therapeutic method is the complete resection of the cyst, regardless of the presence of symptoms that establishes definitive histologic diagnosis, relieve symptoms and prevent complications.
Löffler syndrome by Toxocariasis the treatment with Thiabendazole was started and a significant improvement in patient’s health occurred. Discussion: The toxocariasis is a parasitic disease caused by the larvae of Toxocara spp, mainly by Toxocara canis. Dogs are definitive hosts and contaminate the soil with their feces with eggs of Toxocara. Humans acquire the infection as accidental hosts by ingesting these eggs in contaminated soil. The larvae spreads to the liver, lung, eyes, heart, and brain and can be eliminated by the immune system causing eosinophilic granuloma or remain latent for a long period of time. The symptoms are variable and depend on the quality of immune response of the host and the parasite quantity in circulation. Diagnosis is based on clinical findings of patient with laboratory tests showing leukocytosis, eosinophilia and hypergammaglobulinemia. There is a strong relationship between this parasitic disease and allergic disorders such as asthma. The parasites cause allergic reactions because Larval antigens stimulate Th2 pattern. The differential diagnosis between asthma and toxocariasis with pulmonary involvement is not easy. Many signs and symptoms end up overlapping, which causes a real barrier for correct diagnosis.

188 EVALUATION OF THORACOABDOMINAL DYNAMIC IN THE PRUNE BELLY SYNDROME
José Maria González Neta; Anniele Medeiros Costa; Patrícia Fernandes Barreto Machado; Débora de Araújo Barroso de Pinho; Tânia Wobiel Folescu; Aline Mota Fleming; Instituto Nacional da Mulher, Criança e Adolescente Fernandes Figueira - FIOCRUZ.

Objective: Assessment of the thoracoabdominal dynamic in children diagnosed with Prune Belly syndrome. Case report: Male infant with meconium aspiration syndrome, born at term with 4635g. CPR was conducted in the delivery room. The baby was admitted to the intensive care unit for assisted ventilation with nasal CPAP for 72 hr. Imaging tests showed bilateral hydrenephrosis and lung hypoplasia. He underwent bilateral pieloctomy with 1 Day of life. He was evaluated in 4/2/2013 for computerized evaluation of thoracoabdominal dynamics at the pulmonary physiology lab. The evaluation was conducted before feeding the newborn, who was at deep sleeping and without sedation. The thoracoabdominal movements were analyzed by inductive plethysmography (Respirate). The exam showed significant distortion of the rib cage with phase angle of 170 degrees. Discussion: Examination of thoraco-abdominal dynamics can be useful in assessing the indication of auxiliary mechanisms for optimization of thoracoabdominal movements such as the elastic thoracoabdominal binder.

189 PEDIATRIC BRONCHOSCOPY, EXPERIENCE OF A SINGLE CENTER OF PEDIATRIC THORACIC SURGERY
Angelica Maria Barba Rueda (Programa de Pós-Graduação em Ciências Pneumológicas. Universidade Federal do Rio Grande do Sul – UFRGS); Débora Quiróptero Fernandes (Hospital da Criança Santo Antônio, Santa Casa de Porto Alegre); Julio Oliveira Espinol (Programa de Pós-Graduação em Ciências Pneumológicas. Universidade Federal do Rio Grande do Sul – UFRGS); Cristiano Feijó Andrade (Universidade Federal do Rio Grande do Sul – UFRGS)

Introduction: Bronchoscopy is an invasive and safe tool in the structural and functional evaluation of the upper and lower airways, in suspected cases of congenital malformations of the tracheobronchial tree, pulmonary infiltrates and lung infections in children. Its indications arise with the need to respond to the symptoms and radiological abnormalities in such patients. Objectives: To evaluate the main indications of bronchoscopy in pediatric patients from the Santo Antonio children Hospital, between the years 2006 to 2010, performed by a pediatric thoracic surgeon team. To evaluate complications of pediatric bronchoscopy. Methods: We analyzed all bronchoscopies performed at the Santo Antonio Children Hospital from January 2006 to December 2010 in patients younger than 18 years. Records with incomplete data were excluded. Results: One thousand and four hundred and eighty-eight procedures were included, 813 (55%) were performed in males. The main indications for these procedures were the evaluation of anatomy and dynamics of the airways in 40% of patients, collection of lower airways material in 466 (32%) patients, atelectasis in the chest x-ray in 193 (19%) patients, stridor in 154 (15%) and recurrent pneumonia in 124 (12%) patients. The most frequent endoscopic finding was hyperemia of lower airway (17%), followed by laryngomalacia in 196 (11%) patients and Tracheomalacia in 182 (11%) patients. The exam was normal in only 7% of the procedures. The collection of bronchoalveolar lavage was performed in 1023 (76%) procedures. During this period only 10 patients (0.7%) had some type of complication, being the most frequent one transient hypoxemia (0.14%). Conclusions: The pediatric bronchoscopy is a safe procedure with few complications and can be performed on patients of any age.

190 CONGENITAL CYSTIC ADENOMATOID MALFORMATION OF THE LUNG. A CASE REPORT IN A PRESCHOOL CHILD
Vanessa Dias de Souza Cambraia (Residente em Pediatria do Hospital Municipal Dr. Mario Gatti); Fabricio da Silva Castillo (Residente em Cirurgia Geral do Hospital Municipal Dr. Mario Gatti); Rafael Spinola Cambraia (Médico da Emergência do Complexo Hospitalar Prefeito Edvaldo Orsi); Luiz Henrique Pereira (Cirurgião Pediátrico do Hospital Municipal Dr. Mario Gatti).

This discuss the case of a male pre-school 2 years old with the diagnosed Congenital Cystic Adenomatoid Malformation (CCAM) of the lung and its clinical implications. The child started with a fever, cough and dyspnea for 3 days. Previous medical history of asthma diagnosed one year ago and pneumonia one month ago treated in home with amoxicillín. The respiratory sounds decreased in front and side region of the left hemithorax. The Rx hypotransparency in left hemithorax with mediastinal shift to the right. Diagnosed with pneumothorax held the thoracic drainage and the mediastinal return to original position. However, continued with radiological alteration of the left hemithorax. After completion of the chest ct scan showed massive, gasososa and cystic image of the left without pleural effusion, compatible with (CCAM) of the lung. After segmetectomia the front portion of the left lung apex performed the macro and microscopic analysis of the fragment, confirming the diagnosis. This condition is a rare anomaly of the lower respiratory tract, characterized by exaggerated and uncontrolled growth of terminal bronchi, causing disability in the alveoli and formation of cysts of various sizes, impairing respiratory function. The etiology is unknown, but occurs early in pregnancy. The diagnosis can be performed in the neonatal period and in some cases even their disappearance. Around 80% of cases are diagnosed during the first 2 years of life, usually as complications of respiratory distress, pulmonary effusion, spontaneous pneumothorax and recurrent pulmonary infections. The surgical treatment by lobectomy is the therapeutic choice. The presence of cysts in the lungs jeopardize the formation of the respiratory tract leading to pulmonary hypoplasia and its complications and greater predisposition to lung cancer. Although rare is extremely important to their early diagnosis and treatment, thereby reduce the lung damage caused by their uncontrolled growth.

191 SEVERE ACUTE RESPIRATORY FAILURE CAUSED BY THANATOPHORIC DYSPLASIA. THE REPORT OF TWO CASES WITH DIFFERENT CLINICAL DEVELOPMENTS
Vanessa Dias de Souza Cambraia (1); Mayssa Alvarez Zezende (1); Kátia Maria Roqueiro Gomes (1); Patrícia Santinello Migliorini (1); Thiago Araújo Monteiro (1); Roberto Salvador Martins (Coordenador da Residência em Pediatria do Hospital Municipal Dr. Mario Gatti); 1 — Residente em Pediatria do Hospital Municipal Dr. Mario Gatti;
Objective: This report describes two cases of newborns (NB) with intrauterine diagnosis of Thanatophoric dysplasia (TD) and their distinct clinical outcomes for acute respiratory failure (ARF) serious. Result: Both cases were diagnoses with TD during the second trimester of pregnancy by ultrasonography performed during prenatal care with the following findings: increased nuchal translucency measurement above the 95th percentile, shortening the appendicular skeleton, extreme shortening of long bones, extremely narrow chest, clavicle skull and polyhydramnios. Although the two NB presents the same syndromic diagnosis, only one (CAPURRO 36 weeks and 1 day) had since intrauterine life the diagnosis of pulmonary hypoplasia requiring respiratory support soon after birth and evolving to death in 48 hrs. The second (CAPURRO 39 weeks and 6 days) had no diagnosis of intrauterine pulmonary hypoplasia, but presented ARF in childhood and keeping in invasive mechanical ventilation so far. Both had pulmonary hypertension since the beginning requiring high respiratory parameters and pharmacological approaches to control. Discussion: TD is a rare type of neonatal osteochondrodysplasia and high severity and lethality. It is an autosomal dominant disease caused by mutations of the growth factor receptor gene in fibroblasts 3 (FGFR3), located on the short arm of chromosome 4. Responsible for bone disorders with severe shortening of bones of limbs, macrocephaly and shortening the ribcage. The perinatal survival is usually short and the majority of infants die from severe ARF. The narrow thorax associated with pulmonary hypoplasia with the presence or absence of hydrocephalus is the main explanation for this serious respiratory condition. In the literature there are reports of individuals with survival above the neonatal period, but with a poor quality of life, dependent on mechanical ventilation, multiple hospitalizations for infections, neurological deficit, motor and hearing loss.

192 CASE REPORT – TUMOR CELL GERM MIXED IN YOUNG MAN
Joanna Araujo Simoes (Fundação de Medicina do ABC); Nyla Thyara Melo Lobao; Taylor Gerhardt (Hospital Araujo Jorge); Anete Sevcovic Gramach (Fundação de Medicina do ABC); Arteiro Queiroz Menezes (Fundação Hospital Adriano Jorge).

Objective: The objective of this study is to report the case of a young patient of 16 year-old male resident inside the Amazon, with chest pain complaint a year ago, with previous trips to the emergency room undiagnosed. Results: COB, 16, male, born and raised in Sangal-AM, was admitted with chest pain complaints of pain and tightness in constant left hemithorax for a year, which worsened the physical effort. It evolved with nocturnal dry cough, postprandial vomiting, appetite loss and cessation of walking. It was diagnosed with pulmonary tuberculosis and pleural effusion. Performed thoracentesis relief and rifampicin and isoniazid. Physical examination revealed pallor and emaciated. The throat had bulging of the left hemithorax and left pneumonectomy. Histopathology revealed a germ cell tumor, mature teratoma. The patient was discharged with outpatient treatment and left pneumonectomy. Objective: To report the case of a patient with chronic respiratory failure, secondary to bronchiolitis obliterans in need of prolonged mechanical ventilation, followed by tracheostomy due to failure in weaning from mechanical ventilation attempts and pulmonary lobectomy for organ fibrosis. He was discharged at 1 year and 5 months and started home care since. Today patient is 5 years old. We aim to show the familiar difficulties of patient management tracheostomy and success in their clinical course with home care. Method: The information was obtained through review of medical records, interviewing the parents of the patient, photographic register of diagnostic methods to which the patient was submitted and review of the literature. Final considerations: the case reported and raised publications shed light on the discussion about the treatment and handling of a critically ill patients with poor socioeconomic conditions, a resident of frequently burned area, that despite all the difficulties it succeeded in the course of treatment due to the employee and treatment options, often multi-home monitoring and therapeutic methods agreed not showing satisfactory results with regard to symptomatic relief, reduction in the number of exacerbations of disease and improvement of quality of life.

194 SWEAT TEST: A DESCRIPTIVE ANALYSIS IN A REFERENCE CENTER
Althea Guimarães Faria Guimarães Faria (Department of Pediatrics and Department of Medical Genetics, Faculty of Medical Sciences, State University of Campinas); Fernando Augusto de Lima Marson (1); Antônio Fernando Ribeiro (1); Carla Cristina Souza Gomez (1); Maria de Fátima Servidoni (1); José Diceu Ribeiro (1); 1 – Department of Pediatrics, Faculty of Medical Sciences, State University of Campinas)

Objective: To perform a descriptive analysis of the sweat test performed in a reference center. Material: There are analyzed 5627 exams realized in our reference center. Methods: There was performed a retrospective study of 5627 exams and descriptive analysis of the test including: (i) weight sweat, (ii) age of solicitation, and (iii) chloride values considering the cystic fibrosis (CF) diagnosis. Results: Of the total sample, 2997/5627 (53.3%) were male. The age of the subject at the time of the test was 0 to 6 months – 645/5567 (11.4%), between 6 months to 18 years – 3855/5567 (68.3%) and older than 18 years – 1067/5567 (18.9%). The weight of sweat in the collection was 50 ± 46 (50 to 96) mg. The results for the chloride have been grouped and the values ?found were: 0 to < 30 mEq/L – 3668/5644 (65%), ≥ 30 to < 40 mEq/L – 655/5644 (11.6%), ≥ 40 to < 60 mEq/L – 667/5644 (11.8%) and ≥ 60 mEq/L – 654/5644 (11.6% – CF diagnosis). There was difference when comparing the individual’s age at diagnosis with the results of the sweat test performed for all the groups (p < 0.001). For the comparison considering the positive values for the sweat test and the negative result with the subject age, the odds ratios of 1.467 (95% CI: 1.161 to 1.835), 0.315 (0.266 to 0.373) and 3.208 (2.736 to 3.898), respectively for age groups, 0 to 6 months, between 6 months to 18 years and older than 18 years were observed. Conclusions: The groups 0 to 6 months and older than 18 years had a higher proportion of CF diagnosis. Our sample is representative when compared to other centers.

195 QUALITY OF LIFE AND RESPIRATORY EXACERBATIONS IN PATIENTS WITH CYSTIC FIBROSIS
Monica Ninet Rodas Gonzalez (Hospital Universitário Professor Edgard Santos); Luizy Nathália Teixeira Rocha (1); Carolina de Oliveira Augusto (1); Vinicius Ramos-Machado (1); Almério de Souza Machado Júnior
Pediatric Pulmonology
Describe the quality of life among adults patients with (CF) and verify its relationship with the number of exacerb in the last year. Patients aged 18 years or older with a confirmed diagnosis of CF which agreed to participate clinically stable who responded to the Cystic Fibrosis Questionnaire-Revised (CFQ-R). Meth Cross-sectional study with adult patients recruited from the CF outpatient clinic at a tertiary hospital. QOL was measured using the CFQ-R, a disease-specific health-related quality of life (HRQOL) quest for CF patients that measures functioning in a variety of domains including Physical Functioning Vitality Health Perceptions Respiratory Symptoms Treatment Burden Role Emotional and Social Functioning. Max score corresponds to the highest QOL and minimum score corresponds to the lowest QOL. Number of exacerb was recorded and subsequently categorized as ≥ 1 and ≥ 2 exacerb. Continuous variables were summarized as means and sd and % were calculated for categorical variables. The Mann–Whitney U test used to compare the groups with ≥ 1 and ≥ 2 exacerb and other continuous variables p < 0.05 indicated statistical significance. Results 47 patients of wich 61.7% women and 61% not white skin color; mean(sd) age of 45.0(19) and mean(sd) FEV1 % predicted of 62.4(28.9). Good scores in Body Image. Eating and Digestion domains were observed when analysing CFQ-R with their respective means(sd): 79.6 ± 25.7, 75.8 ± 26.7, 82.8 ± 17.9. When the median values of QOL domains were compared respectively to groups of exacerbations, statistical significance was only found in Vitality, Health Perceptions and Social Functioning domain, as follows: 62.5(50;77.9) vs 50 (33;36,6) and p = 0.01; 66.7(55,6;88,9) vs 55,6(33,3; 66,7) and p = 0.02; 63,9(50; 83,3) vs 44,4(38,9;66,7) and p = 0.02. Reported good scores in Body Image. Eating and Digestion domains. The most exacerbated group (>2 exacerb the last year) presented worse scores in the vitality, perceived health and social domains.

TUBERCULOSIS IN INFANT: AN IMPORTANT DIFFERENTIAL DIAGNOSIS TO BE THOUGHT

Objective: Report a case of a 11 month-old baby with a history of repeated pulmonary infections treated like pneumonia, who got better just when received tuberculosis (TB) treatment. Case report: Female patient, 11 month-old, previously healthy, eutrophic, without neonatal complications. At 8 month-old she was diagnosed with community acquired pneumonia. After treatment, despite being asymptomatic, due to persistence X-ray images, she was treated with amoxicilin-clavulanate and then clarithromycin. However, she presented with fever and knee pain and was hospitalized with the diagnostic of pneumonia and septic arthritis. After discharge, the patient had fever again and was readmitted with diagnosis of infected atelectasis. She was 10 month-old at that time and she still presented cough. There was contact report on 2 occasions with a person diagnosed with pulmonary TB. Chest CT scan showed pulmonary consolidation in middle and lower lobes, tuberculin skin test was 16 mm and Acid-Fast Bacilli detection in gastric sample was negative. Compared to point-scoring system of public health guideline from Brasil to diagnose children aged ten years old and younger, the diagnosis of pulmonary TB in this case was very possible. Treatment with isoniazid, rifampicin and pyrazinamide was started. After the begging of treatment patient had clinical improvement and presented radiological resolution after 4 months of treatment. Discussion: According to World Health Organization at least 550,000 children suffer from TB each year, of which 70–90% has pulmonary disease. TB illness in children is often missed due to non-specific symptoms and difficulties in diagnosis. Children with TB are usually paucibacilar, for this reason microbiology confirmation is difficult. TB remains a major public health problem and should be remembered as a differential diagnosis in cases of children, especially children under 5 years old, with lung disease that does not improve with conventional treatment.

IMPACT OF PASSIVE SMOKING IN ASTHMA SYMPTOMS IN PATIENTS OF A RESPIRATORY OUTPATIENT CLINIC

Objective: To evaluate the impact of passive smoking (PS) on asthma severity. Material and Methods: The study was conducted in 2013 and the sample consisted of asthmatic patients between 3 and 14 years, who were monitoring the respiratory outpatient clinic. A self-administered questionaire was developed by the researchers, in order to assess the impact of PS on asthma severity. It was used the chi-square test to compare the responses of groups with different variables and was considered significant values of p < 0.05. Results: The sample consisted of 77 patients, of among 50.6% were male and 41.6% reported PS at their homes. Those who were PS, most had no symptoms compared to those who had no contact with smokers. It was also observed that the number of crisis, trips to the emergency room and sleep interruption was greater in those who were not PS. On the other hand, there was the increased use of inhaled corticosteroids and leukotriene modifiers in PS. The factors faced with patients who had contact with household smokers, such as controlled asthma were not statistically significant. Discussion: There was not statistically significant between PS and asthma symptoms. Although many epidemiological studies have investigated a possible association between exposure to cigarette smoke and...
allergic diseases in childhood, the epidemiological evidence is still not firmly established. Conclusion: It is necessary to expand dissemination of anti-smoking measures to reduce the prevalence of those patients who passively inhale cigarette smoke, especially in children.

199 TUBERCULOSIS (TB) IN INFANT: RARE CASE WITH EARLY ONSET AND WITHOUT INDEX CASE

Cinthya Covessi Thom de Souza; Universidade Estadual de Maringá-PR

Objectives: TB is a hard disease diagnosis in childhood, because of non-specific symptoms and difficulty to obtain bacteriological confirmation. The objective is to describe an unexpected case with early onset symptoms without contacts. Case: 3-months-old girl with a history of 4 admissions. The first one was at 40 days of life because of fever of unknown origin. Subsequent admission was for febrile conditions associated with wheezing and cough. Treatment was performed hospitalized. She came to our hospital in the third admission with the same respiratory symptoms. An opacification in the right lung was noted and treated as pneumonia. After 3 weeks, began with fever and had persistence of radiological image. Thus, we started the diagnostic investigation. All close contacts were asymptomatics and had negative investigation for TB. She had failure to thrive in the last 6 weeks, HIV negative and immunological status was normal. Tuberculin skin test (TST) < 5 mm. CT scan showed hilar lymphadenopathy with lung infiltrates and some reticulonodular opacities. We performed gastric aspirate (3 samples) and found 2 with smear microscopy and culture both positives for M. tuberculosis. The treatment was done successfully with RHP. Conclusions: The most common presentation of primary pulmonary TB is lymphadenopathy with parenchymal lesions. Although, this infant start with the symptoms with just 40 days old and any index case was found. Congenital TB hypothesis does not fit since the mother had no symptoms of pulmonary or extrapulmonary TB and congenital disease tends to be more severe. Accordingly to the WHO, the diagnostic yield of a set of 3 gastric aspirates is only about 25–50% of children with TB disease. However, we found 2 positives samples of gastric aspirate, confirming the bacteriological diagnosis of TB. Despite rare, primary TB should be kept in mind as a potential diagnosis for infants who are unresponsive to pneumonia treatment.

200 CHILD BRONCHIECTASIS: FROM ETIOLOGY TO DIAGNOSIS CASE REPORT AND LITERATURE REVIEW

Gabriela Pimenta Coelho de Castro; Vinicius Barros Figueiredo; Universidade de Franca

Bronchiectasis is an abnormal and irreversible dilatation of one or more bronchi that derive from other explanations of stasis of secretions, giving possible material of contamination, followed by infection, inflammation, destruction of the bronchial wall that weakens and dilates. Among the various etiologies we find acute infections by virus, bacteria and fungi, asthma, immunodeficiency, cystic fibrosis, enzyme deficiencies, malformations, congenital syndromes (Mounier-Kuhn, Williams-Campbell, Kartagener), aspiration syndromes, dyskinesia Riparian Primary, bronchiolitis obliterans, allergic bronchopulmonary aspergillosis (ABPA), among others. The clinical scenario is heterogeneous and can cause few symptoms even very serious syndromes. It usually has an insidious onset characterized by chronic cough with or without sputum, recurrent infections, hemoptysis, fatigue and anorexia. Diagnosis is mainly by high resolution tomography and other efforts can be done in order to elucidate etiology. Treatment will depend on an underlying cause having, antibiotic prophylaxis as an important weapon. The objective of this study is to report a bronchiectasis scenario in pediatric patients attended in a Basic Health Unit in the city of Franca, making a brief review of the literature on the pathophysiology, main etiologies as well as diagnostic research and practice adopted. The patient of the study has been monitored with antibiotic prophylaxis and is stable. BAAR was performed on three samples of gastric lavage, immunosuppression tests in sweat, chloride test and esophageal serigraphy and all of them were negative. The test results associated with the fact that the patient has been asymptomatic for almost a year and the new tomographic images that show a partial regression of bronchiectasis lead us to believe that possibly these alterations may be a consequence of previous pulmonary infections presented by the patient.

201 PLEURAL TUBERCULOSIS IN ADOLESCENT

Eugenio Fernandes de Magalhães; Marcus Vinicius Landim Stori Milani; Ana Paula de Oliveira Fernandes; Manuel Goavêa Otero Y Gomez; Isabela de Sousa Pereira; Anna Luiza Pires Vieira; Universidade do Vale do Sapucaí – UNIVAS

Objective: To report a case of pleural tuberculosis in adolescent. Material and Methods: Female patient, 16 years old, resident of Pouso Alegre, MG, student, reports low back pain for two months, which worsened after efforts and with the breath, and lost approximately 4 kg in the same period. Denies fever, cough, asthenia and appetite loss and refers only occasional dyspnea. Reports that her cousin is carrying tuberculosis (TB) and is in treatment for 2 years. On examination, patient is in good general condition, afebrile, and with no change in the various devices and systems. Was performed ultrasound of the chest revealed that 10 ml of fluid in the pleural cavity, which prevented the realization of thoracentesis and tuberculosis skin test reagent, 15 mm. With that, pleural tb was diagnosed taking into account the medical history, family history and the radiological finding. The therapeutic regimen was done with rifampicin, isoniazid, pyrazinamide and ethambutol for two months and then isoniazid and rifampicin for another 4 months. Discussion: TB infectious disease is a significant health problem in Brazil, which was ranked as one of the twenty countries in the world with the highest incidence rates of the disease. Pleural TB is historically considered a primary manifestation resulting from hematogenous or lymphatic spread from a pulmonary focus being, childhood TB, different from the adult form, because usually paucibacillary. Diagnostic confirmation is mainly based on the identification of Tb agent in pleural fluid culture or tissue fragment. It points out that in recent years, ADA dosage has been an alternative for the diagnosis of pleural tuberculosis. Conclusion: Considering the high incidence rates in our country, it is essential that doctors in primary care network and pediatricians know the different forms of presentation of the disease in children and adolescents.

202 PULMONARY ABSCESS

Eugenio Fernandes de Magalhães; Marcus Vinicius Landim Stori Milani; Ana Paula de Oliveira Fernandes; Manuel Goavêa Otero Y Gomez; Isabela de Sousa Pereira; Rodrigo de Almeida; Universidade do Vale do Sapucaí – UNIVAS

Objective: To report a case of pulmonary abscess. Material and Methods: Male patient, 10 years old, resident of Congonhal, previously higid, began fever board. Initially was diagnosed with tonsillitis, receiving amoxicillin/ clavulanate. The patient developed intense pain in the left hemithorax and was taken back to service. The examination revealed no fever, regular general condition, pale. In respiratory auscultation observed decrease murmur at the base of the left hemithorax. Remaining unchanged devices and systems. Chest x-ray showed in the lower left third, the presence of cavity with fluid level, suggesting abscess, which was later confirmed by chest computed tomography. Negative blood culture. Started hospital treatment with ceftriaxone and oxacillin. It evolved with the framework of improvement and was discharged in 10 days. Discussion: Pulmonary abscess is a collection of necrotic and supplicative tissue in the parenchyma, the aspiration is the most common cause. It may also be secondary to pneumonia. The most common symptoms are fever, productive cough, chest...
pain, asthenia and occasionally elimination of fetid purulent sputum. The diagnosis is based on clinical presentation, identification of predisposing conditions and radiological changes. The lesions are usually unique, appearing on chest radiography as irregular wall cavity and fluid level inside; chest computed tomography allows better anatomical definition of pulmonary lesions in case of diagnostic uncertainty and is more accurate for the differential diagnosis of empyema. Conclusion: Most cases respond properly to clinical treatment (antibiotics and postural drainage), but sometimes it is necessary any surgical procedure, particularly in cases where there is associated pleural empyema. Rapid assessment and aggressive and early intervention would appear to reduce morbidity, mortality and costs associated with health care.