

**Background and Aims** Informed consent underpins medical practice. However, the immediacy of PICU can make obtaining informed consent challenging. Nevertheless, consent could be obtained for less urgent procedures and investigations.

We aimed to survey the current practice of obtaining consent for commonly performed procedures and investigations.

**Method** We conducted a survey of UK PICUs and Paediatric Transport Services to determine if consent is obtained and the form it takes.

**Results** We obtained responses from 16 PICUs or transport services. No unit has a written consent policy.

#### Abstract 470 Table 1

Procedure	Written Consent	Verbal Consent	No Consent	Comments
Intubation	0	2	14	
Arterial or Central Line	0	2 (including 1 occasionally)	14	Occasionally (n=1)
Blood transfusion	1	1	14	Time permitting (n=1); Occasionally (n=1)
Chest drain	1	2	13	
PD catheter	3	0	8	Not applicable (n=5)
Bronchoscopy (on PICU)	1	1	9	Not applicable (n=5)
Bronchogram	2 (including 1 occasionally)	0	5	Not applicable (n=9); Occasionally (n=1)
CT Scan	1 (only if contrast required)	1	14	Follow local guideline for transport service (n=2)
MRI	4	2	8	Follow local guideline for transport service (n=2)

**Conclusion** Current practice is variable both across and within units. Predictably, consent is most commonly obtained for less urgent procedures; consent may be impossible for time critical procedures. There are aspects of consent that the PIC community may need to review - for example the dichotomy of obtaining consent for the same procedure (eg bronchoscopy) being undertaken in theatre but not on PICU.

#### 471 CARE FOR THE CHILD WITH A RARE DISEASE: A JOINT VENTURE

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Novel digital communications and data registration add to a collaborative management approach for chronic diseases. Primary health care (PHC) providers, patients, and families can effectively prevent many major contributors to the disease burden. Patients need information, motivation, and skills in prevention and self-management. Prevention measures should be provided by professionals. This study aims at establishing opportunities for PHC to detect children with rare and chronic conditions and provide PHC with tools for personalized prevention for children.

**Methods** 931 newborns were followed in the Dutch youth health records, during 2 years to detect children with a rare condition. The preventive scheme includes registration of pregnancy and delivery information, neonatal and vision screening, growth-, development and physical evaluation.

**Results** 12 children had been diagnosed because the child's obvious congenital anomalies recognized at birth or presenting with an

acute illness. In another 10 children, the first signs and symptoms were recorded at the PHC. Of all 22 children, 5 children have a condition suitable for a coordinated care program with help of e health. We developed a system based on clinical "Detailed Clinical Models", for self management and continuous preventive care for children with a rare condition in primary care.

**Conclusion** Rare diseases can be detected in primary care and after proper diagnosis primary care can again be involved in collaborative management.

#### 472 THE INFLUENCE OF AEROPOLLUTANTS ON THE DEVELOPMENT OF ASTHMA (BA) IN CHILDREN

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**Aim** The aim of our study was to evaluate the impact of aeropol- lutants on the development of asthma in children.

**Materials and methods** To solve this problem we used the method of compatibility of geochemical and medical data using GIS technologies and the R-factor analysis. Using our own and compilation data we have created database with the frequency of asthma disease in children in different cities of Crimea. Average annual data on the pollution of the cities by different gases and dust were taken at the local medical service. Then, having made the database with the calculated values of the factors at each point.

**Results** The study in some cities we have found out only 3 global integrated pollution zones. We have found out that the highest incidence of asthma is observed in adolescents and it is 10 times higher than for 7–14-year-old children, this group of children is the most susceptible to the effects of polluting factors. All the studied pollutants: carbon monoxide, dust, nitrogen dioxide, sulfur dioxide, formaldehyde et al. in case of increasing of their levels in the air, are fully involved in the development of asthma in children. But even single limit doses of dust and ammonia (normal maximum allowable doses a year) within the year for children with asthma are significant and contribute to the development of disease exacerbation.

**Conclusion** The R-factor analysis can solve many problems for the prevention of diseases including asthma.

#### 473 DETERMINING THE CORRELATION BETWEEN THE SCAR SIZE OF BCG AND SUFFERING FROM ASTHMA IN CHILDREN, BY ASSESSING THE LEVEL OF THELPER1/ THELPER2

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The Bacille Calmette-Guérin (BCG) vaccine is used more than any other vaccines in the world, whereas a lot of studies have spoken about allergy and its relationship with the BCG vaccine. The aim of this study was to determine the relationship between the levels of Thelper1(INF-gamma)/Thelper2 (IL-4, IL-13) in children suffering from Asthma and the scar size of BCG in them. 100 children who have got scar, 60 of them suffered from asthma and 40 did not suffer from it, were studied. The Chi-square test revealed that the frequency of cases with scar size larger than 5mm, is 43% in Asthmatic patients and 70% in control group that the observed difference was statistically significant ( $P = 0.01$ ). T test also revealed that the average of INF-gamma is considerably lower in patients suffering from Asthma than the control group. ( $6.95 \pm 3.83$  (pg/ml) in asthmatic group compared with  $10.75 \pm 6.98$  (pg/ml) in control group) ( $P = 0.001$ ) whereas the average of IL-4 ( $30.90 \pm 16.51$  (pg/ml) in asthmatic group compared with  $9.95 \pm 7.44$  (pg/ml) in control group) ( $p < 0.001$ ) and IL-13 ( $48.85 \pm 13.66$  (pg/ml) in asthmatic group

compared with  $10.49 \pm 12.44$  (pg/ml) in control group) ( $p < 0.001$ ) is higher. It was demonstrated that the average of INF-gamma/IL-13 ( $0.3054$  in asthmatic group compared with  $1.9334$  in control group) in patients suffering from asthma, is lower. The size of BCG scar in Asthmatic patients was, significantly, smaller than the control group. Moreover, the average of Thelper1 (INF-gamma) was lower and the average of Thelper2 (IL-4.13) was higher in asthmatic patients. Therefore, there is a correlation between the size of BCG scar and the levels of Thelper1 (INF-gamma) and Thelper2 (IL-4.13) with asthma; thus, there could also be a correlation between the scar size of BCG, Thelper1 and 2.

#### 474 HUMORAL IMMUNITY IN CHILDREN WITH CHRONIC TONSILLITIS

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**Background** Chronic tonsillitis is a common problem for pediatricians and otorhinolaryngologists.

The aim of our study was to evaluate and compare the humoral immunity in children and adults with chronic tonsillitis.

**Subjects** We examined 24 children and 13 adult patients with chronic tonsillitis.

**Methods** Immunologic examination included analysis of: total leukocytes; absolute and relative lymphocytes; C reactive protein (CRP), antistreptolysin O (ASO), rheumatoid factor; investigation of humoral immune factors: absolute and relative B lymphocytes, serum IgA, IgM, IgG, circulatory immune complexes, interleukin amount - IL-4, IL-8, IL-1 $\beta$  in blood serum.

**Results** Increasing level of the absolute lymphocytes amount ( $2.92 \pm 0.24$ ;  $2.2 \pm 0.11$ ) ( $p = 0.04$ ), interleukin - 8 ( $229.09 \pm 73.63$ ;  $35.05 \pm 14.64$ ) ( $p = 0.047$ ) and interleukin - 1  $\beta$  level ( $191.19 \pm 68.44$ ;  $10.88 \pm 4.28$ ) ( $p = 0.045$ ) ( $p = 0.045$ ) were observed in children. Average indices of interleukin-8 and interleukin - 1  $\beta$  in children is in 6.5 and 17.4 times higher than in adults.

Serum IgA level in adults was significantly higher than in children ( $1.36 \pm 0.16$ ;  $1.92 \pm 0.19$ ) ( $p = 0.038$ ). An antistreptolysin-O titre in adults in comparison to children ( $162.5 \pm 45.53$ ;  $216.67 \pm 45.78$ ) is higher in 1.4 times.

**Conclusions** Nonspecific defense factors play an important role in children with chronic tonsillitis, while in adults the main role is held by specific immune response. Comparative analysis of anti-inflammatory cytokine synthesis by immune competent cells in chronic tonsillitis proved the fact that in children it is much higher than in adults.

#### 475 AUTOIMMUNE LYMPHOPROLIFERATIVE SYNDROME IB: IMPROVEMENT WITH RAPAMYCIN

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**Introduction** Autoimmune lymphoproliferative syndrome (ALPS) is a disorder on a defect in the apoptosis of lymphocytes with linfo-proliferation and immune dysregulation. Type Ib, defined by mutation in the gene that encodes the protein FAS-ligand, is a rare entity.

**Case report** A 9-month-old male infant was referred because a failure to thrive and abdominal distention. Physical examination showed pallor, signs of severe malnutrition, axillary and inguinal lymph nodes, hepatomegaly and giant splenomegaly. Family background: parents, cousins of Moroccan origin, brother died at the age of 4 when he was under study for a giant visceromegaly. Complementary tests highlighted severe anemia (Hb 7.30 g/dl) and thrombocytopenia (platelets 76000/mm<sup>3</sup>), paravertebral lymphoid

proliferation of 4 cm, elevation of IL10, soluble CD25, 28% T lymphocytes double negative (CD4 - and CD8-) in peripheral blood and cell culture with defect of apoptosis in one of the samples. The genetic study identified the mutation of the gene TNFSF6 which encodes FAS-ligand-protein. After the diagnosis of ALPS type Ib, treatment with rapamycin was started at doses of 2 mg/m<sup>2</sup> diary. An optimal evolution was observed, with a reduction of visceromegaly size after 30 months of treatment, without adverse events by the time.

**Conclusion** We emphasize the importance of the suspicion of this entity in children with chronic visceromegaly, especially with family history. Despite the few existing data on treatment with rapamycin for this disease and children in general, we have seen an appropriate response and a good tolerance in this patient.

#### 476 A CASE REPORT OF KARTAGENER'S SYNDROME ASSOCIATED WITH NASAL POLYPS

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Nasal polyps are benign nasal masses that can cause nasal obstruction, headache and snoring. The overall incidence or prevalence of nasal polyposis is unknown. They are diagnosed more frequently in men and during the third and fourth decades of life. Nasal polyps are rare in children. Most clinical data indicate that there is no greater prevalence of nasal polyps among atopic compared with normal populations.

A 14-year-old male patient was referred by an otolaryngologist to investigate the cause of nasal polyposis. There were otitis media, sinusitis and bronchial asthma to patient's history therefore he used inhaled corticosteroid for control long term asthma, sometimes bronchodilator to asthma attack and antibiotic treatment for sinusitis. No family member had similar respiratory complaints or any significant systemic illness.

His physical examination was found above 97% for weight and height difficulty breathing through nose, bilateral nasal polyposis, rarely sibilant ronchi on pulmonary auscultation and his heart sound was heard deeply. Laboratory findings; Hemogram was normal, IgA:189mg/dl, IgE 203 IU/L, inhalen sIgE and epidermal prick test were negative, Chest X-Ray showed dextrocardia, A computed tomograph scan of the chest showed situs inversus with lateral segment of right middle lobe tubular bronchiectasis, peribronchial thickening and atelectasis, the right lower lobe tubular bronchiectasis. Pulmonary function testing demonstrated a mixed obstructive and restrictive pattern.

The pathogenomic findings of Kartagener Syndrome are dextrocardia, bronchiectasis and sinusitis. His findings were consistent with Kartagener Syndrome. This patient were presented due to nasal polyposis associated with Kartagener Syndrome and delayed diagnosis.

#### 477 CHRONIC RECURRENT SEVERE LIP ANGIOEDEMA IN YOUNG CHILD SECONDARY TO ALLERGIC RHINITIS

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**Background and Aims** Allergic rhinitis (AR) is a significant disease that affects children and adults and often under-diagnosed which has a deleterious impact on quality of life. Chronic recurrent severe lip angioedema secondary to AR was not previously reported.