

PO-0359 HOW TO SELECT THE SWEAT TEST CANDIDATE. 10 YEARS OF EXPERIENCE IN SCREENING FOR CYSTIC FIBROSIS IN CHILDREN

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Background Sweat test (ST) remains gold standard in cystic fibrosis (CF) diagnosis. Alarm symptoms are age-related.

Aims Retrospective review of cases subjected to ST.

Methods Patients were selected by paediatricians, neonatologists, surgeons, based on suggestive symptoms, personal (PH) and familial history (FH). Inclusion criteria: for 0–1 month age group, patients with PH of atelectasis, meconium ileus, intussusception; 1–12 months, recurrent wheezing (RW), failure to thrive (FTT); 1–5 years, previous group symptoms, plus chronic cough/diarrhoea; >5 years, 1–5 years symptoms, plus recurrent pancreatitis/sinusitis. For all age, patients with PH of salty taste of sweat (STS), salt wasting syndrome, heat shock (HS), and FH of CF, azoospermia. ST was performed with Nanoduct system. Values <60 mmol/L (equivalent NaCl) were considered normal, between 60–80 equivocal, >80 positive. Normal values patients were eventually retested, those with equivocal at least once, those with positive mandatory twice.

Results Were performed 406 ST (344 patients): at 0–1 month 11 tests (45,4% peritonitis), at 1–12 months 175 (65,5% RW), at 1–5 years 108 (25,9% FTT), at >5 years 50 (62,5% repeated pneumonia-RP). 4 tests equivocal, 5 false positive, 6 positive (5 infants: 1 RW, 1 HS, 2 atelectasis, 1 STS; 1 male 4 years old RP). Relating to age, only 1 positive from 141 RW infants (p 0,03) and 1 from 15 RP children; 2 positive from 6 atelectasis infants (p 0,0001), 1 from 2 HS and 1 from 2 STS infants (p 0,0002). Neither pancreatitis, FTT or positive FH.

Conclusions Great attention on infants with other symptoms than classical ones: higher statistical significance for STS, HS, atelectasis.

PO-0360 BONE MINERAL DENSITY IN EGYPTIAN CHILDREN WITH FAMILIAL MEDITERRANEAN FEVER

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Background Familial Mediterranean fever (FMF) has episodic or subclinical inflammation that may lead to a decrease in bone mineral density (BMD).

Objective To assess BMD in Egyptian children with FMF on genetic basis.

Subjects and methods A cross sectional study included 45 FMF patients and 25 control children of both sexes, with age range between 3–16 years old. The patients were reclassified into 2 groups: **Group I (A)** 23 cases used colchicines for 1 month or less, and **Group I (B)** 22 cases used colchicines for more than 6 months. For both patients and control, MEFV mutations were defined using molecular genetics technique and BMD was measured by DXA at 2 sites: proximal femur and the lumbar spines.

Results four frequent gene mutation where found in the patient group: E148Q (35.6%), V726A (33.3%), M680I (28.9.0%) and M694V (2.2%). There were also 4 heterozygous gene mutations in 40% of control children. Patients received Colchicines

treatment for less than 1 month had highly significant lower values of BMD at femur and lumbar spines than control children ($p < 0.007$, $p < 0.001$). Patients received Colchicines treatment for more than 6 months had improved values of BMD at femur compared to control, but there were still significant differences between them at lumbar spine ($p > 0.036$). There are insignificant effect of type of gene mutation on BMD and the risk of osteopenia among the patients.

Conclusion FMF had significant effect on BMD. However; regular use of colchicines treatment improves this effect mainly at femur.

PO-0361 FOODBORNE ILLNESSES IN MOROCCAN CHILDREN: RESULTS OF A NATIONWIDE STUDY

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Background and aims Foodborne illnesses are a serious public health issue, especially among children in developing countries. The aim of this study is to determine the magnitude of foodborne diseases and associated risk factors among children in Morocco.

Methods We conducted a retrospective study of foodborne diseases in children under the age of 6 years, notified between 1992 and 2008, to the Moroccan Poison Control Centre.

Results There were 2 589 cases of food poisoning and 12 deaths reported in children during the study period. The foods most often involved were dairy products, fish and fishery products. The median delay in presentation to hospital was 9 h. The majority of children had gastrointestinal symptoms, associated or not with respiratory, cardiovascular and neurological symptoms. On admission to hospital, the majority of patients received symptomatic treatment. According to the results, region of origin, types of symptoms and the delay before treatment were significant risk factors influencing patients' health status. The risk of death due to poisoning was significantly higher among patients presenting with arrhythmias, apnea, hypothermia, hypotonia, bradycardia and coma.

Conclusions Foodborne diseases are a significant public health problem in both developed and developing countries. The effective treatment remains prevention focused on public awareness about the dangers of food poisoning, especially for young children.

PO-0362 THE SILVER-RUSSELL SYNDROME: REPORT OF 2 CASES

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Background The Silver-Russell syndrome is rare pattern of malformations which associated growth retardation generally starting in antenatal period, a characteristic facial feature and limb asymmetry. We report two new cases illustrating this syndrome.

Observation 1 A female infant was admitted in paediatric department at the age of 4 months. The neonatal period was unremarkable; the infant had no intrauterine growth retardation (birth weight and height ranging between the 25th and 50th percentile). The infant's head circumference was large contrast with

small facial mass. Clinical findings included a small triangular face with frontal bossing, blue sclerae, thin lips with downturned corners, retromicrognathia, and limb asymmetry. Weight and height at admission were less than 5th percentile (Weight was 3.9 kg and size 56 cm); her head circumference grew normally along the 75th percentile.

Observation 2 A male Infant was hospitalised at the age of 1 year and 4 months. Physical examination showed growth deficiency: Weight: 7.1 Kg (<5th percentile), size: 72 cm, head circumference: 46 cm (25th percentile). The same facial features as the first case was found with body asymmetry, flat and narrow feet and cryptorchidism.

Conclusion The diagnosis of this syndrome is based on clinical findings. The treatment is purely symptomatic and growth hormone administration does not achieve the target size. The long-term prognosis, a part the growth, is good.

PO-0363 ASKIN TUMOUR:CASE REPORT

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Introduction Askin tumour is a primitive neuroectodermal tumour of the thoracopulmonary region. Pulmonary location is not common. We report a case of an Askin tumour in a child who presented with worsening breathlessness.



Abstract PO-0354 Figure 1

Observation A 9 years-old female child with no pathological history, was admitted with severe dyspnea up to one week. A Weight loss was reported. Physical examination showed silence in pulmonary auscultation of the left lung field. A chest X-ray found a left opacity occupying almost the entire of the left lung, mediastinal compression to the right side and nibbled aspect of the fourth rib. Chest Ultrasound showed pleural effusion with finely heterogeneous liquid. Chest CT showed a pulmonary solid tumour with aspect of massive necrosis of the left upper lobe with hemothorax, rib erosion in contact, without lymphadenopathy or vascular invasion or secondary lesion distance. A scano-guided biopsy was performed and immuno-histological examination found a small round cell tumour with morphological aspects favour the diagnosis of primitive peripheral neuroectodermal tumour corresponding to an Askin tumour. The child underwent an intensive chemotherapy and surgery.

Conclusion The management of Askin tumour is not well codified. The prognosis of these tumours remains very unfavourable because of their metastatic risk and potential local recurrence.

PO-0364 HISTOLOGICAL ASPECTS OF THE FETO-PLACENTAL INTERFACE IN THE PREGNANCY INDUCED HYPERTENSION

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Abstract PO-0354 Figure 2