

seven Palestinian RHS patients (tyrosinemia type 11) diagnosed based on serum tyrosine levels and other clinical manifestations of the disease. DNA sequence analysis and BLAST were used to search for abnormality in the TAT gene after sequencing the 12 exons and exon-intron boundaries included within the gene. Two mutations were identified; a nonsense mutation (R417X) in two RHS patients (previously reported in a French Patient), and a splicing mutation (T408T) (previously reported in two Palestinian brothers) in the other five patients. Moreover, six polymorphisms could be identified, three were previously identified including IVS11 +143 a > g, IVS8 +113 t > c, and S103S and three new ones that include g→t @-17, IVS7+84 c > g, IVS7-73 g > t. The T408T splicing mutation appears to be specific to the RHS Palestinian families as none of these nucleotide transversions were reported in other populations. These results provide the bases for implementing molecular genetics analysis for precise diagnoses of patients and carriers of the disease in and provides a strong tool for genetic counseling programs within the population. It also provides a powerful tool for prenatal diagnoses of fetuses for parents who are carriers of the disease. These results also provide the bases for selection of specific intron-exon sequences in the analysis process among RHS suspected patients who are referred to the various medical centers and clinics for diagnoses and treatment.

20) *Optimizing Chelation Therapy in Thalassemia patients; Bridging Clinical Data and Patient Management*

Prof. Hisham Darwish, *Faculty of Medicine, AlQuds University, Abu Dies, Jerusalem and Thalassemia Patients Friends Society (TPFS), Ramallah, Palestine.*

Abstract

Patients with thalassemia major who receive regular blood transfusions are likely to develop iron overload. This will result following saturation of the iron carrying capacity of transferrin, which generally takes place after 20 transfusions. Consequently, excess plasma iron (labile iron) is cleared rapidly by the liver, heart and endocrine tissues at a rate that exceeds 200 times normal uptake of transferrin-bound iron. Excess labile iron within cells will destroy the structure and function of mitochondria, lysosomes, lipid membranes, proteins and DNA. The clinical consequences can include liver cirrhosis and fibroses, cardiomyopathy, diabetes and other endocrine disorders. With proper control of body iron at all times in patients, these effects are preventable but only some are reversible once tissue damage has occurred.

The primary rate of iron chelation therapy is to bind and remove iron from the patient body at a rate either equal to or greater than the rate of iron uptake of transfused iron. Complete chelation should aim to achieve both iron balance and iron detoxification. In patients who accumulated dangerous tissue iron levels, removal of this iron is also highly desirable. Several iron chelators have been developed to help achieve these objectives. The recent introduction of the effective oral chelator deferasirox provides chelation coverage and significant control of LPI levels over the entire 24 hour period. Evidently, administration of the drug at a dose of 20-30 mg/kg/day significantly controls iron levels in plasma, liver and myocardiocytes in thalassemi a major patients. Accumulating clinical data from key studies indicate that thalassemia patients can lead a happy and enjoyable normal life similar to normal individuals if their treatment is performed right and they receive the proper clinical attention from the medical staff.

Definitely, chelation therapy requires close monitoring of patients vital organ status and the treatment protocol should be adjusted accordingly. Long term treatment with iron chelation highlights the importance of titrating the dose of the drug for each patient according to individual rates of iron intake from continued blood transfusion, current iron storage levels, safety markers (renal function for example) and target body iron content desired.



The presentation will highlight some of the recent clinical data from leading clinicians in chelation therapy from several parts of the world and the lessons that can be learned from their experience and work for the benefit to optimize the treatment of thalassemia major patients in our population.

21) Contamination of baby milk "The Chinese story"

Mohammed Jawad Musmar, Ph.D,R.Ph, *Dean, College of Pharmacy, An-Najah National University*

Infant formula industry is an 8-10 billion dollar per year business. Across the globe, huge advertising budgets are spent to convince women that it is better and more convenient to bottle feed their babies.

Advantages of breast milk over formula milk have been extensively studied, however in this paper issues of contamination will be explored.

Although mother milk may contain more dioxins, PCB,s,and organochlorine pesticides than infant formula, health concerns for milk formula may include the risk of contaminated water, potential contaminants in bottles and nipples, and contaminants in the formula itself.

In most parts of the world, water is polluted with microorganisms, chlorine byproducts, weed killers, insecticides, solvents, lead ,and arsenic.

Formula itself may have contaminants introduced in the manufacturing process such as broken glass fragments, Salmonella, fungal toxins that may cause cancer. Again metals like Aluminum, manganese, cadmium had been detected.

The list also includes high level of plant derived estrogens or genetically modified organisms, antibiotic residues, diesel fuel from trucks and several pollutants from packing.

Is the plastic baby bottle safe? which type of plastic is safe? Is bottled water safe?

Most recent issue of infant formula safety is that "Chinese baby milk formula is contaminated with melamine". What is melamine and why milk is contaminated? What other baby food products imported may be contaminated? Can we test our products for pollutants including melamine?

The presentation will cover all issues related to milk formula safety.

22) Blood lead level among school children, a developed method for blood lead measurement

Ahed H. Zyoud, *Department of Chemistry, An-Najah National. University*

Abstract

Lead and it's compounds are used as additives to several products such as gasoline and paints. Lead has a toxic effect especially on brain and nervous system. Almost no published work has been found providing information about blood lead levels in children of Palestine. And thus the present work was carried out.

An improved (ASV/HDME) method for determination of lead in whole blood by anodic stripping voltametry (ASV) using hanging dropped mercury electrode "HDME" has been developed with a special reagent at An-Najah N, University laboratory.

A total of 518 sample from 10th grade students of Jenin district, the samples have been analyzed using the mentioned improved method>

The geometric mean of blood levels was 87.75 µg/L, a variation in the geometric mean of blood lead level was noticed with respect to students place of residence;(Camp student 119.43 µg/L), (City students 92.41µg/L), (Yammon village students 77.65 µg/L). also a variation appeared with respect to

