NEWS AND VIEWS

NEWSLETTER FROM PUERTO RICO

In Puerto Rico hematologic investigations went on actively in the year 1957. They dealt with the following subjects:

Sprue: “A Ten-Year Study of Synthetic Folic Acid as a Nutritional Supplement and Therapeutic Agent” by Spies, Suárez, García López and Stone1 stresses the necessity of folic acid for the metabolic processes of cellular life. It is a progress report of a cooperative study which was begun in 1945 and which is still being continued. Primarily it is an appraisal of the work initiated by the authors in the Caribbean area showing that folic acid and vitamin B12 are useful in combating sprue and nutritional macrocytic anemia. From the beginning this group has been interested in the absorption and utilization of food, and they have learned that the patient treated with these effective nutrients (vitamins) eats much greater quantities of food than before therapy. The authors describe the response of the first cases of sprue and nutritional macrocytic anemia given a synthetic therapeutic agent—how they changed from severely ill people to people who were able to work and who, 10 years later, were still in excellent health. Practicing physicians in Puerto Rico have applied the principles recommended in these studies, with the result that it is virtually impossible to find severe “virgin” cases of sprue or nutritional macrocytic anemia in Puerto Rico. The authors have shown that in the prevention of disease or, if one prefers, in the maintenance of good health over a period of a decade, folic acid like other important vitamins has the greatest usefulness.

E. Pérez Santiago and collaborators have continued their studies on sprue. They consider this disease one of the conditions due to or associated with the so-called malabsorption syndrome. They reported on the various absorption tests including the xylose, vitamin A and butter fat tolerance tests.

Charles E. Butterworth, Jr. and E. Pérez Santiago2 reported this year (1957) in Boston on jejunal biopsies in sprue. Histologic studies were performed on jejunal biopsies obtained at laparotomy from 8 to 10 inches from the ligament of Treitz in 6 patients with sprue. Edema, inflammation and widening of the villi were observed in all cases. The role of faulty epithelial regeneration was discussed as a possible etiologic mechanism for the histopathology observed.

It may be of interest to note that Milanes and collaborators3 from Cuba reported on jejunal biopsy findings in a case of untreated sprue in 1951. They also reported shortening or thickening of the villi and a lymphoplasmocytic infiltration in both the epithelium and the lamina propria and called attention to thickening of Auerbach’s plexus and degenerative changes in neurons.

Butterworth and Pérez Santiago suggested the desirability of continued studies with jejunal biopsies using the small bowel biopsy tube described by Shiner in England or the one introduced on this continent by Crosby.

Thrombotic Thrombocytopenic Purpura: Rodriguez, Babb, Pérez-Santiago and Costas-Durieux4 reported the case of a 36-year-old woman suffering from thrombotic thrombocytopenic purpura who, practically moribund, recovered after splenectomy and is still alive one year after the operation.

Hemoglobin: R. Menéndez Corrada5 at the 1956 meeting of the Puerto Rico Medical Society reported on a hemoglobin survey with filter paper electrophoresis conducted among 500 unselected new admissions to San Patricio Veterans Administration Hospital during the years 1955–1956. Of this number, 13 patients (2.6 per cent) were found to have abnormal hemoglobins. Eight of the 13 showed the combination AS, while there was one instance of pure S (sickle cell anemia). The total percentage showing S hemoglobin was 2.5 per cent. One single case of hemoglobin S- Hb C disease was found, the first reported in Puerto Rico. Another case also exhibited the combination SC but was ap-
parently healthy. Finally, two cases of AC were found. The total hemoglobin C percentage incidence was 0.8 per cent.

Mercedes V. Torregrosa, A. Rivera Trujillo and Oscar Ruiz Soler* reported on abnormal hemoglobins among Puerto Ricans and concluded:

1. The incidence of sicklemia among 616 Puerto Ricans was found to be 4.21 per cent. Among persons considered Negroes, 6.7 per cent had the sickle cell trait.

2. In the family studies of 12 patients originally diagnosed as suffering from sickle cell anemia, hemoglobin-C was found in one of them, associated with hemoglobin-S. The rare combination of sickle cell spherocytosis was present in another. In addition, a case of thalassemia of intermediate severity was diagnosed in Puerto Rico for the first time in a child of Greek racial extraction.

3. Six of the remaining patients had clinical, hematologic and electrophoretic findings of classical sickle cell anemia. Electrophoretic studies were very helpful in elucidating the final diagnosis in three of their patients; these had been diagnosed as suffering from sickle cell anemia, whereas only a sickle cell trait was found. The signs and symptoms present in these patients were attributed to other nonrelated disease entities.

**Hemophilia and Hemophilia-like Syndrome:** Mercedes V. Torregrosa and Angel Clintrón Rivera' reported on 17 patients who presented a hemophilia-like syndrome. The laboratory findings pointed towards a deficiency of thromboplastin as the cause of the hemorrhagic diathesis. Thirteen patients were males and had a family history of bleeding; eight were classified as classical hemophilia or hemophilia-A. Studies to identify the type of hemophilia could not be performed in five.

Hemophilia B or PTC deficiency was found in one male patient with no family history; the disease appeared to be congenital since it started very early in life. A female patient with acquired Christmas disease (PTC deficiency) was also studied; this case will be published in detail because the authors believe it represents a condition that has not yet been reported.

Circulating anticoagulants (anti-thromboplastic in nature) were found in two female patients (these two cases have been published in Blood).

**B-Glucoronidase in Hypersplenism with Hemolytic Anemia:** Busó, Olavarrieta and Suárez* reported on the possible role of B-glucoronidase as an important chemical factor in the production of hemolysis in hypersplenism.

They studied a young adult with hemolytic anemia and marked splenomegaly due to intestinal schistosomiasis. Intracorpuscular factors to account for the hemolytic process were absent: The patient's Hgb was type A on filter paper electrophoresis, and the apparent Cr^½ half-life of the red cells, which approximated 2.7 days in the patient's circulation, was over 30 days in a normal recipient. Circulating extracorpuscular hemolytic factors were repeatedly found lacking: no circulating agglutinins and/or lysins were found at different temperatures or on decreased plasma pH values, and the direct Coombs' test was repeatedly negative.

The authors studied the rate and degree of hemolysis occurring in vitro when washed erythrocytes were incubated for 24 hours at 37 C. after treatment for 20 minutes with a freshly prepared suspension of B-glucoronidase containing 500 Fishman units per 100 ml. of inactivated normal AB serum or normal saline solution. The control erythrocytes showed no hemolysis during the initial 12 hours and a maximum total of 6 per cent hemolysis in 24 hrs. The patient's red cells, on the other hand, always started to lake before the 12th hour, and total hemolysis was invariably present long before the 24th hour. Partial inhibition of this process to a total range of 65 per cent to 85 per cent hemolysis was achieved by varying concentrations of cortisone acetate.

Splenectomy under corticosteroid medication resulted in complete clinical and hematologic recovery. The hemolytic pattern in vitro was duplicated postoperatively by substituting splenic tissue fractions obtained by Fishman's method instead of B-glucoronidase. The main pathologic finding was congestion of a markedly increased splenic “red pulp.”

A two-year follow-up examination revealed no hematologic abnormalities in the patient. His red cell apparent Cr^½ half-life was 27 days. More interesting perhaps, his
erythrocytes behaved like the normal controls when they were again subjected to the hemolytic action of B-glucoronidase in vitro.

The Anemia of Hypothyroidism: Roberto Búso et al. of Fundación de Investigaciones Clínicas performed the following studies:

The blood of 11 adult hypothyroid patients was labeled with radioactive chromium. The red cell affinity for radiochromium in vitro and the apparent Cr half-life in vivo was determined and found to average 54.5 per cent and 13.8 days, respectively. These figures are at variance with the normal values of 80 per cent and 24 days previously established in this laboratory. The frequency of anemia in hypothyroidism is easier to understand if we conclude that the erythrocytes have a shortened life span.

Blood Copper in Normal and Pathologic States: These studies, performed at the Fundación de Investigaciones Clínicas by R. M. Suárez and collaborators, Santurce, and reported at the regional meeting of the American College of Physicians, October 26, 1957, showed that although there are small but high-grade copper deposits on the island, and although there is apparently no lack of this element in the soil, in some of the pasture grasses and in a few of the fresh waters of Puerto Rico, the blood level of copper in apparently healthy Puerto Rican people was lower than that reported for continental U.S.A.

In a series of 217 healthy subjects studied, there were 35 (16 per cent) with blood copper values below 50 micrograms per cent. They found, as others have elsewhere, that whole blood copper levels are higher than serum or plasma levels, and that women showed higher blood copper values than men. They also found that the age group between 40 and 49 years showed lower blood copper values than other adult age groups.

Whole blood copper values were found to be 84.0, 89.8, and 83.6 μg. per cent for men and women; while serum copper levels were 76.6, 79.8, and 81.8 μg. per cent for men and women.

Copper values were found to be lower in cattle than in pigs, sheep, and goats. The mean levels were 54 μg. per cent for cattle, 164 μg. per cent for pigs, and 134 μg. per cent for sheep and goats.

In pathologic states they found hypercupremia in Hodgkin's disease and in chronic leukemia. In hypochromic anemia, as well as in pernicious anemia, normal values for copper were obtained in remission and in patients suffering from peptic ulcers, but rather low levels were found in patients with sprue in relapse.

Although in the medical literature hypothyroidism is included among the conditions in which hypercupremia may occur, all eight cases of myxedema studied showed definite hypocupremia. No case of nephrosis or hepatocellular degeneration (Wilson's disease) was studied.

The authors could not account for the high incidence of hypocupremia among healthy Puerto Ricans, except on a nutritional basis. The beef from local slaughter houses, as well as some of the potable water, might be low in copper, and the richest sources of copper such as nuts, dried legumes, dried fruits and leafy vegetables such as cauliflower and celery, are rarely, if ever, consumed by the native Puerto Rican.

Blood Proteins: In a paper that appeared in the Boletín de la Asociación Médica de Puerto Rico in April, 1957, Suárez, Olavarrieta, Búso, Suárez, Jr. and Sabater from Fundación de Investigaciones Clínicas reported their studies on blood proteins in healthy Puerto Rican adults and in various animal species. They made a plea for standardization of paper electrophoresis and proposed the biuret-albumin equivalent conversion factor (BAE).

The data obtained were in general similar to those reported from other American and European laboratories where similar methods were employed.

There was more gamma globulin and less albumin in the older than in the younger group of humans, as well as in other animal species studied.

According to their estimates, 1 per cent albumin as obtained directly from the scanner represents 96 mg. of albumin by the biuret method, with a standard deviation of 85 mg. to 108 mg., and in the globulin fractions 1 per cent is equivalent to 60 mg. with a standard deviation of 59 mg. to 61 mg.

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REFERENCES


VIIth CONGRESS OF THE INTERNATIONAL SOCIETY OF HEMATOLOGY

At the VIIth Congress of the International Society of Hematology, which was held in Rome in September, 1958, under the Presidency of Professor G. Di Guglielmo, two Ferrata Foundation awards (Premio Ferrata) were given, one to Dr. William Dameshek of Boston, the other to Dr. Maxwell NI. Wintrobe of Salt Lake City. The awards were made at different plenary sessions by Professors Introzzi of Pavia, President of the Ferrata Foundation and Professor Di Guglielmo at ceremonies during which diplomas and checks (for 500,000 lira*) were presented. This was followed by a lecture by the award recipient. Dr. Dameshek's subject was "The Myeloproliferative Syndromes with particular reference to Erythremic Myelosis (Di Guglielmo Syndrome)," and Dr. Wintrobe spoke on "The Changing Aspects of Clinical Investigation. A Study of Acute Leukemia."

The Ferrata Foundation awards are made in honor of the memory of the late great Italian hematologist, Professor Adolfo Ferrata.

At the same Congress, the Henry and Lillian Stratton Foundation award was presented by Dr. med. (Hon.) Henry M. Stratton to Professor Ludwig Heilmeyer of Freiburg, Germany in the form of a Paul Ehrlich Medal and a check for $500. Because of Dr. Heilmeyer's absence due to an acute illness, the award was accepted by his assistant Dr. Begemann. Ehrlich medals were also presented to the previous award winner Professor Jan Waldenström of Malmö, Sweden and to the Presidents of the VIIth and Vth Congresses, Drs. Di Guglielmo and Dameshek respectively.

At the official Banquet of the Congress, Dr. Moises Chediak—in very moving terms—presented magnificent plaques representing Cuban hematology to Drs. Di Guglielmo and Dameshek, as Presidents of the present and the just previous Congresses.

This highly successful VIIth Congress of the International Society of Hematology was held in the magnificent halls of the "Esposizione Universale di Roma" with approximately

*About $800.
2000 participants from all parts of the world. Plenary and multiple sessions were held for six days, with one half day being spent at an audience with the late Pope, Pius XII. The 1960 Congress will be held in Tokyo and that in 1962 in Mexico City under the Presidency of Professor Ignacio González Guzmán.

BOOKS RECEIVED FOR REVIEW


