*editorial*
*Haematologica 1996; 81:385-6*

# IDA BIANCO SILVESTRONI. FROM RED CELL OSMOTIC FRAGILITY TO MOLECULAR BIOLOGY OF GLOBIN GENES. A LONG, PRODUCTIVE LIFE WITH THALASSEMIA

This issue of Haematologica contains an interesting paper by Ida Bianco (Figure 1) and co-workers on the molecular diagnosis of α-thalassemia,[1] one of the innumerable contributions (by no means the last) that this gentle, inexhaustible lady has made to our present knowledge of thalassemia. I met her last spring in Naples, of course at a meeting on thalassemia, and, like the rest of the audience, I was fascinated by her up-to-date competency and enthusiasm.

Ida Bianco was born in Rome on July 30, 1917, and received her M.D. from the University of Rome School of Medicine in 1941. While doing post-doctoral work with Dr. Ezio Silvestroni (later her husband) (Figure 2), she decided to reproduce some experiments by a Czech investigator, Hoffman, who had described a decrease in osmotic fragility in some patients with cancer (presumably because of the associated iron deficiency anemia). Bianco and Silvestroni employed the Simmel test in 50 cancer patients and 50 normal controls. They found that 2 patients and 4 controls presented a clear decrease in osmotic fragility and erythrocytosis at the same time. Based on these preliminary findings, they decided to use a modified Simmel test (the Silvestroni-Bianco red cell fragility test) for a screening of the normal population.

Within a group of 400 normal individuals, 7 were found to have decreased osmotic fragility, anemia, microcytosis, hypochromia and additional red cell morphological abnormalities. Most importantly, this condition was shown to be hereditary since in the 3 cases studied at least one of the parents was shown to carry the same disorder. On November 26, 1943, Bianco and Silvestroni presented their work on *Microcythemia* as an oral communication at a meeting of the Rome Medical Society. They also recognized that microcythemia shared many similarities with the Rietti-Greppi-Micheli syndrome (hemolytic jaundice with decreased red cell fragility).

Figure 1. Ida Bianco Silvestroni in 1996.

Figure 2. The late Ezio Silvestroni.

Thomas Cooley had described his series of cases of splenomegaly in children, with anemia

Figure 3. Drs. Ida Bianco and Ezio Silvestroni performing their screening tests for β thalassemia.

and peculiar bone changes (β thalassemia major), in 1925, but scientific communication between the US and Italy was difficult in those years. Nonetheless, Bianco and Silvestroni rapidly realized that their disorder was the heterozygous state of Cooley's anemia. In 1946, in fact, by studying several families from the Ferrara region they were able to establish this relationship clearly. In 1944, they also reported the first case of microdrepanocytic disease (sickle cell-β thalassemia).

Between 1945 and 1960 Bianco and Silvestroni carried out fundamental screening studies in Italy (Figure 3), showing that β thalassemia was a social problem in this country with about 2,500,000 heterozygous individuals and over 10,000 homozygous patients.

In 1952, they created the Center for Studies on Thalassemia in Rome, which is still active under the direction of Dr. Ida Bianco. Every working day Dr. Bianco applies her screening tests (osmotic fragility, red cell morphology and red cell counts) to some hundreds of students from Lazio (the region around Rome). She personally examines peripheral blood smears and it takes 10 to 20 seconds for her to identify the morphological abnormalities of *Microcythemia.* This program, supported by the Lazio Regional Administration, has been particularly fruitful since there has not been a single new case of β thalassemia major in this region in the last few years.

In this issue, Dr. Bianco and coworkers present a rational approach to the diagnosis of α thalassemia based on a simple PCR methodology. In most instances, heterozygous α thalassemia is still suspected – by excluding iron deficiency and β thalassemia in a patient with microcytosis – but not confirmed using a specific test. Dr. Bianco's approach fills this gap.

We are grateful to Ida Bianco for her major contributions to our present knowledge of thalassemia and for the tremendous work she has accomplished for the diagnosis of thalassemia syndromes and their prevention. Haematologica is looking forward to receiving further outstanding manuscripts from this evergreen physician.

*Mario Cazzola*
*Haematologica Executive Editor*



***References***

1. Foglietta E, Deidda G, Graziani B, Modiano G, Bianco I. Detection of α-globin genes disorders by a simple PCR methodology. Haematologica 1996; 81:387-96.