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Iron malabsorption and hypochromic anemia in a case of Turner's syndrome

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Summary. Refractory hypochromic anemia was investigated in an adult with Turner's syndrome. Reduced iron absorption, serum iron, and iron incorporation were found in association with increased iron binding capacity and plasma iron turnover. 14 of 57 additional XO subjects were found anemic.

A 30-year-old woman with a karyotype demonstrating X monosomy and various clinical stigmata of Turner's syndrome had been followed for intestinal malabsorption for several years without anemia. In 1977, she developed a mild hypochromic anemia (hemoglobin nadir 11.0 g/dl), which was unresponsive to oral iron treatment. Further evaluation revealed defective iron (^{59}Fe) absorption on 3 serial determinations (range: 0.2–0.3%, low serum iron, (range: 9.8–13 µmole/l; normal 14–31 µmole/l), elevated total iron binding capacity (36 µmole/l; normal = 18–30 µmole/l), and normal reticulocyte counts. Ferrokinetic studies demonstrated a markedly shortened plasma iron turnover (140 mg/day/100 ml of whole blood) and borderline low iron incorporation (62.2%). The latter finding was found in association with an abnormal Shilling tests (3.9% B_{12}-^{59}Co; 6.1% B_{12}-^{57}Co with intrinsic factor). A radiographic examination of the gastrointestinal tract demonstrated abnormal mucosal contours of jejunum and distal ileum, but was otherwise normal.

Normally iron balance is precarious in female subjects, but relatively few clinical entities are commonly associated with iron malabsorption[1]. The most important of these malabsorptive states is gluten-sensitive enteropathy (GSE), although abnormalities in gastric, pancreatic, and biliary secretion have been proposed as additional causes, none has been found clinically important[1]. The late and insidious onset of anemia in the propositus might be explained on the basis of a longstanding generalized state of intestinal malabsorption. It is apparent that iron absorption studies may be the most sensitive indicator of GSE and other generalized states of intestinal malabsorption and anemia often occurs only at an end stage in such conditions[1].

Although ferrokinetic studies have fallen somewhat out of fashion, because the half-time disappearance curve of plasma iron, plasma iron turnover, and percentage utilization provide only an approximation of the state of erythropoiesis; such studies remain a valuable and rational approach to the assessment of anemic subjects[2]. Despite the inherent limitations of the methodology, it is likely that the increased plasma iron turnover and marginal iron incorporation in the propositus may be a response to her chronic iron and vitamin B^{12} malabsorption. Usually the bone marrow in B_{12} deficiency responds dramatically to the anemic stimulus, but because of the inability of the erythroid precursors to produce nucleic acids normally, the erythropoietic effort is largely ineffective; resulting in an increased PIT but a reduced iron incorporation and ultimately, anemia. However, the relationship of these findings to the underlying chromosomal abnormality remains to be elucidated in Turner's syndrome.

Consultation with colleagues and a thorough review of the medical literature, assisted by a computerized system of medical data retrieval (Medlars), yielded no previous reports of abnormal ferrokinetic data in Turner's syndrome. In reviewing our patient records, we found 14/57 (24.6%) anemic subjects with XO karyotypes. In 10 cases, the anemia was mild (hemoglobin $> 10 < 12$ g/dl) and moderately severe in the remainder (hemoglobin $> 8 \leq 10$ g/dl). 2 cases in the latter category were iron responsive. One of these was associated with gastrointestinal telangectasia. Although the frequency of anemia in this series of Turner's syndrome patients does not exceed that expected in a general population of individuals of this age and sex distribution[3], it should be emphasized that iron deficiency anemia is much less common among young women in Sweden, where iron supplementation of foodstuffs is governmental policy[4]. Further investigation of ferrokinetics in Turner's syndrome may be fruitful, particularly in subjects with microcytosis, anemia or malabsorption.

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