# CONGENITAL FACTOR VII DEFICIENCY PRESENTING AS IRON DEFICIENCY ANAEMIA — CASE REPORT

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## INTRODUCTION

CONGENITAL Factor VII Deficiency was described by Alexander in 1951 (Wintrobe, 1974, Hardesty, 1974). By 1964, 40 more cases were added. Approximately 70 cases of Hereditary Factor VII Deficiency have now been reported (Wintrobe, 1974). The inheritence is autosomal, heterozygotes having a partial deficiency and homozygotes having more severe deficiency (Marder, 1964).

Both sexes are affected equally. While certain haemorrhagic manifestations in this condition are shared by both sexes, epistaxis and haemarthrosis have a male predilection while menorrhagia is severe enough to cause signs of iron deficiency (Owen, 1964). We report such a patient who was in cardiac failure as a result of iron deficiency due to menorrhagia.

## CASE REPORT

Patient, an 18 year old student, admitted with a history of about week's duration of dyspnoea, swelling of legs and loss of appetite. She bled excessively during her periods in the last three years. Apart from mild haemarthrosis and epixtaxis, she had no history of bruising either spontaneously or after a trauma. Nor any history of prolonged bleeding after a cut or after dental extraction was elicited. She did not have any malaena or haematuria. Her diet seemed adequate. There was no history suggestive of diseases of the cardiovascular system, renal or liver.

She had an elder brother who had swelling of the knee which subsided spontaneously. She had three other brothers and sisters who do not give any history suggestive of haemostatic disorder.

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Examination revealed extreme pallor, angular stomatitis, loss of papillae of the tongue, and oedema without jaundice. There were no bruises or petechial haemorrhages. Both fundi showed haemorrhages. Lymphadenopathy, hepatosplenomegaly or sternal tenderness was absent. Hess's test was negative. There was evidence of congestive cardiac failure.

The provisional diagnosis made was of iron deficiency anaemia due to menorrhagia and confirmed by laboratory investigations. Laboratory investigations which confirm the diagnosis of iron deficiency anaemia were carried out. There include the haematological indices, blood film, serum iron, total iron binding capacity and bone marrow iron stores.

She was treated for her cardiac failure with frusemide and digitalis. Her iron deficiencies was corrected with packed cells, dose iron infusion followed by oral iron.

In the course of investigations for her menorrhagia, tests of haemostatic functions such as prothrombin time, partial thromboplastin time, clot retraction, clot stability test and thrombin time were done which revealed Factor VII Deficiency. Acquired causes of Factor VII Deficiency were looked into by carrying out liver function tests, the Euglobin clot lysis time, and by looking for LE cells and fibrin degradation products, but was not detected.

Family studies showed one brother had clinical signs of haemarthrosis and two out of five in the family had laboratory evidence of Factor VII Deficiency as indicated by a normal partial thromboplastin time and a prolonged prothrombin time.

## DISCUSSION

Iron deficiency anaemia, the presenting feature in this case, was obviously due to menorrhagia. This had affected the patients for three years

without the cause being determined. Laboratory investigations concluded that the menorrhagia is the result of congenital Factor VII Deficiency. Although rare, it is mandatory on the part of the physician to investigate along these lines in patients with iron deficiency with menorrhagia; less cases of Factor VII Deficiency be missed. Thus more cases may come to light if we make this a routine procedure in the investigation of anaemias. Owen (1964) reported the following percentage of occurrence of the above findings, epistaxis (42%) haemarthrosis (26%) and haematuria (6%).

The patient did not have prolong bleeding after trauma which is in line with Hall's (1964) finding there is paucity of episodes excessive bleeding after trauma or surgical procedures other than dental extraction.

One brother had haemarthrosis. He was found on laboratory investigations to have Factor VII Deficiency. Owen (1964) considered Factor VII of 85% to be normal, 25% — 69% to be heterozygotes and less than 25% to be homozygotes.

Eliciting family history is often a neglected facet of clinical examination. This patient is a case in point to stress the significance of such history taking ritual. So much can be assessed even prior to laboratory investigation.

All the three patients seen by Owen (1964) had anaemia. In these cases, coagulation defects were not initially considered. Since patients may present with menorrhagia the tendency is to attribute the iron deficiency anaemia to menorrhagia. Treating the symptoms of iron deficiency anaemia per se without detecting the actual cause will be of no avail. Again any radical procedures, if contemplated for surgery can be beset with problems of severe bleeding. This can be channelled with care once we know the cause.

## SUMMARY

An 18 year old girl who presented with cardiac failure as a result of Iron Deficiency anaemia due to menorrhagia. She was treated for iron deficiency anaemia until investigation of haemostatic functions revealed Factor VII Deficiency. This disorder is inherited as an autosomal recessive trait. Though clinical evidence of Factor VII Deficiency was found in one member of the family, laboratory investigations revealed deficiency in the other members of the family.

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