# MECHANICS OF EPISTAXIS IN RHEUMATIC FEVER

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Though a link between nose bleeding and rhuematic fever has been suspected for long and inclusion of epistaxis among the minor criteria (Jone Ducket 1944) has been practised for more than 30 years it was not until the 1970s that exploration into the mechanism involved was attempted.

Kovlevo et al 1969 suggested a drop in the three main phases of coagulation without any specifieation. Mirzoyan 1971, reported a decrease in thromboplastic activity, again without specifying if the defect is in the serum, plasma or platelets. He also stressed a lowering of plasma prothrombin activity in the active rheumatic phases, a statement which was contradicated later by Loutfi al 1971.

The present work attempts to throw further light on the illdefined corners of any coagulopathy that may play part in the bleeding occuring from the nose of children with active Rheumatic disease and to specify the defect involved. To achieve this aim the following studies were performed and the results were compared to data obtained from controls.

1. Bleeding time.
2. Coagulation time.
3. Platelets counts
4. Prothrombin time
5. S. Fibrinogen
6. S. Calcium
7. Thromboplastin generation test, using plasma, serum and platelets separately.
8. Histopathological studies of nasal biopsies

The following tables illustrate our main -ve data:

It is clear that there is a defect in thromboplastin generation in the active Rhc phases associated with epistaxis. The defect is observed only when the patients plateles are used, but is absent if patients serum or plasma are used for the test. It can thus be concluded that there is no defect in any of the coagula tion factors present in the sera or plasma of the patient and the platelets number being wihing normal, the defect must be a functional platelets defect.

The functions of platelets in maintaining a normal coagulation process and for preventing a bleeding tendency are manyfold.

Besides containing several coagulation factors which help thromboplastin generation, their aggregating properties and their antiheparin factor also help to hamper any bleeding tendence. The cause of defective platelet function in Rhc Fever is difficult to ascertain. It may well be the result of the autoimune process suggested for the pathogenesis of Rhc Fever.

Worth here to point to the lowered serum phospholipids in Rhc Fever, disclosed by several authors (Kamel 1957, Abdel Khalik 1960, El Miniawi and Abdin 1971). In fact Coburn has suggested that a lowering in lipids is involved in the pathogenesis of Rhc Fever and on this assumption he suggested the egg diet as a treatment of Rhc Fever.

A lowered phospholipid has been linked with a diminished platelet factor 3 activity and would thus be linked indirectly with thromboplastic activity. Whether the lowering in phospholipids is again part of the Autoimmune insult or an inborn defect is not at present established. A role of other biochemical changes in epistaxis is however lacking, serum fibrinogen being actually increased while serum calcium being on high normal levels.

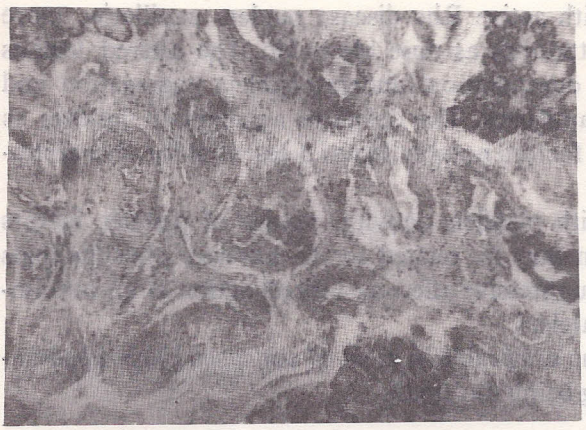
*Histopathological changes seen in active Rhc disease:*

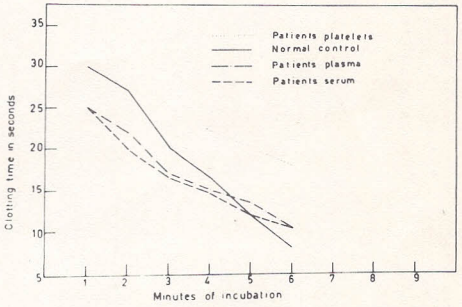
The histopathological finding disclosed in the nasal biopsies were also of particular interest. A specific vasculitis characterised by fibrinoid degeneration was a constant finding in all biopsies obtained during the active Rhc phase. Fibrinoid degeneration was remarkably absent from the control series, with and without nose bleeding. Such fibrinoid degeneration is thus rather specific and probably be of help in diagnosis of atypical Rhc cases. Further it may be a factor in the bleeding diethesis which occurs in Rhc Fever. It is well established that healthy fibrous tissue is important for platelet aggregation and the role of denaturated fibrous tissue in hindering platelets aggregation has been illustrated by several workers.

It is thus highly probable that the vasculitis occuring in the nasal mucasa not only causes it to be more susceptible to traumatisation and oozing but it probably helps indirectly the bleeding diathesis by interfering with platelet aggregation. Such aggregation of platelets is important not only for sealing the oozing capillaries but as well for liberation of platelets coagulation factors.

It is also worth noting the absence of intravascular thombosis in our biopsies, a finding that would be expected in coagulopathies, but which was a matter of controversy among some authors.

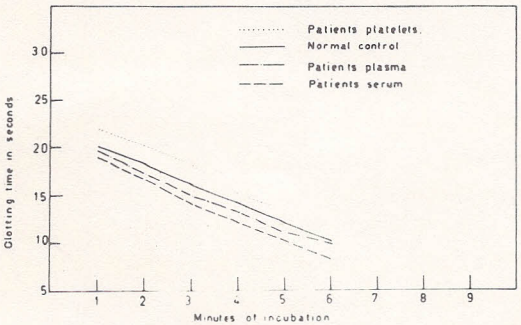
Thus our picturing of the mechanism of nose bleeding in Rhc Fever can be sumarried as such :–






(A) THROMBOPLASTOGRAM OF A CASE RHEUMATIC CORDITIS WITH EPISTAXIS

It is evident that the incubation mixture in which the patients platelets was tested show the longest clotting times allover the period of incubation (4 minutes) which denotes a platelet defect




(B) THROMBOPLASTOGRAM OF A CASE OF RHEUMATIC CORDITIS WITHOUT EPISTAXIS

No defect in the various clotting factors is evident

The vasculitis associated with active Rhc disease renders capillaries more susceptible to ordinary small traumas as nose blows etc. The altered fibrous tissue manifested by fibrinoid degeneration hinders platelets aggregation and the sealing of the ruptured capillaries. The defective aggregation hinders as well the release of coagulation factors notably factor 3 essential for thromboplastic generation.

Other factors interfering with platelets activity, release of platelet factors and thromboplastic generation may be related to the lowered phospholipids in blood and platelets or to a suppressive affect as a direct action of an autoimmune reaction affecting the platelets.

The lowered prothombin time noticeable in most of our cases plays apparently a minor role in the lowering being too small.