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# THE PROBLEM OF THE BLEEDING STATE

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The problem of the patient who bleeds excessively or alleges he bleeds abnormally, is a familiar one. In some cases the bleeding state may be obvious, since purpura, easy bruising or excessive haemorrhage direct one's attention to it. Sometimes when there is a local lesion it may be difficult to know whether to ascribe the bleeding to local or general causes. A patient, for instance, who presents with epistaxis, may have a local lesion in the nose or a generalized bleeding disease, or both these conditions. Patients with haematuria, haematemesis or melaena may present similar problems.
In general there are 2 main groups:

(*a*) *Bleeding State of recent onset*

This normally implies the onset of a new disease, the bleeding being a symptom. Purpura and bruising are the commonest presenting manifestations though bleeding from skin and mucous membranes may also be present. The underlying disease may be one of many but the following conditions occur with considerable frequency:

Idiopathic thrombocytopenic purpura, toxic-allergic purpura (Henoch-Schönlein group), severe blood diseases like leukaemia and aplastic anaemia and disseminated malignant disease. Careful enquiry should be made into possible toxic factors, e.g. drugs the patient has taken (not forgetting the drugs the doctor himself has prescribed) or possible occupational exposure to noxious substances. A careful history should be taken and physical examination done with especial reference to anaemia (out of proportion to blood lost by haemorrhage), lymph-node, spleen or liver enlargement. All these signs might point to possible blood diseases like leukaemia. Bone tenderness is also a valuable sign for which careful search should be made. Examinations of the peripheral blood and the bone marrow examinations may clinch the diagnosis.

Many tests can be done to attempt to find the cause of the bleeding but the most useful one is an assessment of the platelet numbers. If platelets are plentiful on the blood smear, or if no platelets can be seen on a well stained film of peripheral blood, then a platelet count will add little or nothing to the facts already established. If the patient is thrombocytopenic then the bleeding time is generally prolonged, clot retraction and prothrombin consumption are defective, and the tourniquet test will generally be positive. It is rarely necessary to do these tests in patients who have few or no platelets since they are, in the main, merely a manifestation of a thrombocytopenic state.

*Obstetrical patients* present a separate problem since there are a number of bleeding conditions associated with pregnancy. Local causes must always be considered but occasionally the excessive bleeding is due to a haemorrhagic state. One variety may be associated with fibrinogen deficiency. The blood appears to be incoagulable and haemorrhage is sometimes very severe. It occurs not only *per vaginam* but also presents as purpura, a haematemesis and melaena. Without drastic measures to combat the fibrinogenopenia (combined with rapid delivery in most cases) bleeding may become uncontrollable.

(*b*) *Bleeding State present since infancy or early childhood*

Haemophilia (and its variant Christmas disease which can only be distinguished from haemophilia by specialized investigations in the laboratory) is probably the commonest and most serious member of this group. The active manifestations of bleeding occur only in males. The diagnosis can generally be made on clinical grounds. In about 65% of cases there is a family history of bleeding affecting males and being transmitted through apparently unaffected females. (Note: The absence of a family history is not uncommon and does not exclude the diagnosis). There is no other familial bleeding diathesis inherited in this way. Bleeding is always severe, the tendency to bleed being lifelong. Bleeding episodes generally follow trauma, though apparently spontaneous haemorrhage, especially from mucous membranes, is not infrequent. Patients bruise easily but purpura is not a feature. Haemarthrosis is almost invariable at some time or other in anything but the mildest grades of haemophilia, and careful examination will generally disclose ankylosis of one or more joints.

Laboratory examination is necessary to clinch the diagnosis. The coagulation time is usually prolonged but a normal coagulation time can occur in the less severe grades of the disease. This simple test needs to be rigidly standardized. It should be noted that the bleeding time and the prothrombin time are normal; so are the platelet count, clot retraction and tourniquet tests. These patients do not generally bleed excessively from venipuncture or from a needle prick such as is done in measuring bleeding time. The prothrombin-consumption test and the thromboplastin-generation tests are abnormal but these tests should be left to specialized laboratories.

Other congenital or hereditary bleeding diseases are rare. Some, e.g. factor 5, factor 7 or prothrombin deficiency, affect the prothrombin complex and are disclosed by a prolonged one-stage prothrombin time; others are more difficult to diagnose. In some the capillaries are defective. In hereditary capillary telangiectasia abnormal vascular dilatations which bleed can be seen on skin and mucous membranes. Although the disease is present at birth, active bleeding may not occur until the late teens or even the early twenties. The telangiectasia are often well seen on the tongue or on the buccal mucosa. Epistaxis is perhaps the commonest symptom and can be very severe. The diagnosis is obvious on sight. In another group the capillary abnormality is shown by a prolonged bleeding time in the presence of a normal platelet count and normal coagulation time.

There remain a fairly large residue of patients who have mild symptoms of bleeding and in whom no laboratory abnormality can be disclosed. Bleeding may for instance continue for up to 48 hours after tooth extraction without obvious cause. Many women bruise without obvious explanation. Occasional patients aver that they bleed excessively after trauma but no laboratory confirmation can be obtained. Possibly these are physiological variants of 'normal'. Perhaps, as our knowledge extends, we may be able to find the cause of these, at present unexplained, phenomena.

## TREATMENT

As in all diseases diagnosis must always precede therapy. If the patient has a serious blood disease the treatment is as for that disease. Some thrombocytopenic states can be helped by steroid therapy and/or splenectomy, but treatment along these lines should only be considered once the diagnosis is established beyond reasonable doubt. Vitamin K will only aid a proportion of the patients in whom the 'prothrombin' is low. Vitamin K1 is especially useful when the hypoprothrombinaemia is due to drugs of the dicoumarol or phenindione group. Neither vitamin K nor K1 is of any value if the prothrombin is normal. It can be stated dogmatically that calcium is of no value in any naturally occurring bleeding disease. Vitamin C is only of use in the extremely rare case of scurvy.

In haemophilia, treatment is preventive and involves the avoidance of all possible trauma. Major (or even minor) surgery is extremely hazardous. A pint or two of blood is of very little value in controlling the bleeding which occurs in haemophilia. It is to be hoped that one of these days a really potent antihaemophilic substance will become available but, until it does, practitioners would be well advised to handle this problem with extreme circumspection.