The patient I would like to discuss this afternoon demonstrates a problem which is quite common in pediatric practice, namely that of severe and recurrent epistaxis. The patient is a 9-year-old boy who had manifested repeated episodes of epistaxis since the age of 2 years. The nosebleeds occurred most commonly during the night and more frequently during the winter than the summer months. They usually were not associated with trauma although, on occasion, they did follow sneezing. They seemed to occur more frequently when the child had a concurrent upper respiratory infection. In most instances the nosebleeds stopped spontaneously after a few minutes, but on at least three occasions they were so profuse as to require hospital attention and nasal packing.

At the age of 3 years, the child was seen by a physician who informed the mother that something was wrong with the child's platelets and that transfusions would be required each time the child had a nosebleed. During the subsequent 2 years the child was admitted to the hospital for transfusion on several occasions. When the child was 4 years of age the mother sought further advice from a hematologist and the patient was completely evaluated from the standpoint of hemorrhagic disease. No abnormality was found. However, the first physician insisted that a platelet defect was present and informed the mother that transfusions were still required with each nosebleed in order to prevent exsanguinating hemorrhage. Over the succeeding 5-year period the child received numerous transfusions of whole blood, and in recent years with fresh frozen plasma. The nosebleeds had not increased in number or frequency during this period and the child was otherwise clinically well. Two weeks ago the child was brought to the Children's Memorial Hospital for further study.

The past history disclosed no evidence of a bleeding tendency apart from the nosebleeds. Specifically there had been no easy bruising, no bleeding following circumcision, or eruption of the teeth, and no development of hematoma after the usual immunization injections. Complete family history was likewise negative for any evidence of an abnormal tendency to bleed.

On physical examination the child was a well-nourished, well-developed boy who gave no evidence of any chronic illness. The skin was clear and there was no sign of purpura or ecchymosis. There was no glandular enlargement and the liver and spleen were not palpable.

On laboratory examination the complete blood count was entirely normal. The platelets were normal in number and in morphology. The coagulation studies consisted of a bleeding time, clotting time, clot retraction, tourniquet test, prothrombin time, prothrombin consumption and thromboplastin generation tests. The combined sensitivity of these tests was such that even a minor coagulation abnormality would have been detected. All of the coagulation studies were normal.

Thus we have a patient who has been having recurrent and often severe nosebleeds for a 7-year period, and the question of the significance of this symptom is raised. The pediatrician seeing such a patient must consider whether the severe epistaxes constitute evidence of a more generalized hemorrhagic disorder. The problem is often complicated by the fact that many of the children presenting this symptom are just those in whom the question of tonsillectomy and adenoidec- tomy is raised because of recurrent upper respiratory infections. It has been pointed out quite frequently that in the child who does have an underlying hemorrhagic disorder the procedures of tonsil-
Electrolysis and adenoidectomy constitute two of the most dangerous surgical procedures which may be undertaken. The pediatrician, therefore, has several points to evaluate: First, is the child sick? Second, is it necessary that the child be subjected to complete coagulation studies which are not only extensive but expensive? Third, is it justified to proceed with what clinically seems to be indicated, namely the tonsillectomy and adenoidectomy, without intensive study of the child?

In an attempt to evaluate these problems, we have performed complete coagulation studies on 34 children who presented with the primary complaint of recurrent nosebleeds. In each instance, epistaxis constituted the reason for which consultation was requested. The battery of tests listed above was performed in each child.

On the basis of the history and physical examination, it was possible to divide the 34 children into two distinct groups. This is indicated in Table I. In the first group of 17 children, history and physical examination indicated that epistaxis was the only indication of an abnormal tendency to bleed. In other words, the family history was negative and there was no history of easy bruising, of bleeding after dental extraction, circumcision, immunization or, had it been performed, tonsillectomy. In addition, the physical examination disclosed no indication of petechiae or ecchymoses. You will note from the table that no coagulation abnormality was found in any individual in this group. The results of the laboratory findings were confirmed in at least half of the children in this group by the fact that tonsillectomy and adenoidectomy were subsequently performed with no difficulty from excessive bleeding.

In the second group of 17 children, in contrast, further history elicited other indications of an abnormal tendency to bleed, namely, easy bruising, persistent oozing from dental sockets after dental extraction, persistent bleeding after tonsillectomy in those instances where the operation had been performed, and not infrequently a history that other members of the family were also known to bleed excessively. In this group a striking difference in the results of the coagulation studies may be noted in Table I. Here there was only one child who did not have a hemorrhagic disorder.

Of the children with demonstrable hemorrhagic disease, four were found to have PTA (plasma thromboplastin antecedent) deficiency, otherwise known as Hemophilia C. This is a congenital deficiency of one of the plasma thromboplastin precursors which is transmitted as a mendelian dominant and which occurs in males and females. One patient was found to have PTC (plasma thromboplastin component) deficiency, also known as Hemophilia B. This, too, results from a congenital deficiency of one of the plasma thromboplastin precursors but is transmitted as a sex-linked recessive. Of great interest was the fact that 11 of the 16 children were found to have a type of hemorrhagic disorder characterized by a definite vascular abnormality. As shown in Table I, two types of disorders comprise this group. Pseudo-
tility. There is, however, no actual defect in the coagulation mechanism. In vascular hemophilia (affecting seven of the children) an apparently identical abnormality of the small vessels is accompanied by a deficiency of antihemophilic globulin, leading to a dual defect.

Both pseudohemophilia and vascular hemophilia occur in males and females and seem to be transmitted as mendelian dominants. Both are associated with a markedly prolonged bleeding time as contrasted with deficiencies of antihemophilic globulin, PTC and PTA in which the bleeding time is normal. Thus, a fairly simple clinical test was capable of suggesting the presence of a true hemorrhagic disorder in two-thirds of the patients in whom an underlying abnormality actually existed. Unfortunately, however, the other hemorrhagic states which were encountered in this series, namely PTA and PTC deficiencies, demand rather extensive coagulation testing in order to establish the diagnosis. These disorders are associated with a normal bleeding time, a normal prothrombin time and, in all but the most severe instances, with a normal clotting time. Tests such as the prothrombin consumption and thromboplastin generation tests are required in order to detect the deficiencies.

I should like to emphasize as strongly as possible that normal results in the so-called routine coagulation tests, bleeding and clotting time, do not constitute adequate screening for the presence of an underlying hemorrhagic disorder. Children with mild to moderate PTC, PTA and AHG deficiencies, with completely normal whole blood clotting times, may bleed dreadfully if subjected to operation.

The results of this study have indicated to us that if, on careful history and physical examination and family history, epistaxis proves to be the only manifestation of bleeding, it is most likely that the child does not have an underlying hemorrhagic disorder and that an operative procedure may safely be undertaken.

If, on the other hand, examination and inquiry provide additional evidence of an abnormal bleeding tendency in the child or in his family, it is mandatory that complete coagulation studies be performed. In this group of patients the bleeding time may serve as a good clue to the type of disorder which may be present but certainly not all the patients will be detected by this means.

In closing, it must be mentioned that epistaxis, of course, occurs as a manifestation of several systemic diseases such as rheumatic fever, glomerulonephritis, leukemia, sickle-cell anemia, and others. As mentioned earlier, however, the children presented today are referred because of recurrent epistaxis in the absence of any other evidence of systemic illness. It may also have seemed surprising to you that no instances of thrombocytopenic purpura were encountered in this group. In our experience epistaxis has rarely been the sole presenting manifestation of the child with thrombocytopenic purpura.

We believe, however, that a platelet count is one of the basic tests which should be carried out in any child demonstrating any evidence of abnormal bleeding.

**Question:** Dr. Schulman, in those 11 patients who had vascular abnormalities, would the tourniquet test have been positive?

**Dr. Schulman:** In pseudohemophilia and vascular hemophilia the tourniquet test is quite variable and is not positive in more than 50% of the cases. Generally, it is not of much help. If it is positive, it certainly indicates that an abnormality may be present. However, if it is negative, it rules out nothing.

**Question:** What was the incidence of nasal allergy and/or other allergies in the first group of 17 children with no other findings?

**Dr. Schulman:** We were not impressed that allergy played a significant role in the causation of the nosebleeds in this group.

**Question:** In the 17 patients who had no hemorrhagic disease, what was the explanation?

**Dr. Schulman:** That is the most difficult question of all. Most of these children had been seen by otorhinolaryngologists and a great many of them had been cauterized in an attempt to eliminate a seemingly abnormal blood vessel. We were not impressed that any organic abnormality of the nasal vasculature played a significant role in the causation of the nosebleeds. As indicated earlier, the affected children seemed to have a rather high incidence of upper respiratory infections and, in many, obvious hypertrophy of tonsils and adenoids was evident. Other factors leading to nasal congestion, such as dry and overheated rooms in school, picking of the nose, allergy, etc., have been suggested but I must say that by and large the pathogenesis is obscure. An interesting aspect is the rather striking
tendency of those nosebleeds to occur at night while the child is asleep. This may suggest some alteration in circulatory dynamics as one of the possible etiologies.

**Dr. Hsi:*** I would like to ask Dr. Schulman if any of the patients had portal hypertension, either primary or secondary?

**Dr. Schulman:** No.

**Question:** Do these children become iron-deficient?

**Dr. Schulman:** Yes, quite commonly. The nosebleeds may be extremely profuse and blood loss may be appreciable. In the child who has very frequent recurrences of nosebleeds, significant iron deficiency may certainly be encountered.

**Question:** In the pseudohemophilias, did you find evidence of abnormal capillaries?

**Dr. Schulman:** Yes. In all of the cases reported here studies of the capillaries, either in the nailbeds or in the bulbar conjunctiva, were performed. Abnormal capillaries as indicated by marked coiling and tortuosity were seen in all.

**Question:** So frequently, Dr. Schulman, one gets a history from the parent that other members of the family were “bleeders,” and still there is no evidence of hemorrhagic disease. Is that type of history significant?

**Dr. Schulman:** This is quite true and may often be quite misleading. However, we found in this study that the family history may be extremely helpful in evaluating the symptom in the child. For example, despite the statement that other members of the family have been “bleeders,” further questioning will frequently elicit the fact that these individuals have been challenged and have not, in reality, demonstrated a true tendency to bleed excessively. Thus, we always ask whether these “bleeders” have had dental extractions performed or have had tonsillectomies and adenoidectomies in childhood; in women, we inquire about the extent and duration of menstrual bleeding. We believe quite strongly that, if an individual has sustained dental extractions and/or tonsillectomy and adenoidectomy without demonstrating excessive bleeding, this individual does not have one of the congenital hemorrhagic disorders.

It is not infrequent to find a definite family history of epistaxis. The fact that the older members of the family who, in childhood, have also presented with epistaxis, have sustained operative procedures without excessive bleeding, is helpful confirmatory evidence in deciding that the child under study does not have a true hemorrhagic disorder. In general the significance of epistaxis in other members of the family is quite similar to that which we have found for the children. That is, if epistaxis has proven to be the only manifestation exhibited by the adult, no true hemorrhagic disease has been present. In numerous instances we have performed complete studies on the adult members of the family who have given such a history. The conclusions derived from the studies in the children have been confirmed in the studies of their older relatives.

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