The Significance of Epistaxis in Childhood

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lectomy 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-hemophilia (affecting four of the children) apparently results from an inherited anatomic and functional abnormality of capillaries and small arterioles leading to failure of contrac-

### TABLE I

**EPISTAXIS IN CHILDHOOD**

<table>
<thead>
<tr>
<th>History</th>
<th>No. of Cases</th>
<th>Type of Disorder Found</th>
<th>None</th>
<th>PTA Deficiency</th>
<th>PTC Deficiency</th>
<th>Pseudo-hemophilia</th>
<th>Vascular Hemophilia</th>
</tr>
</thead>
<tbody>
<tr>
<td>Epistaxis only</td>
<td>17</td>
<td></td>
<td>17</td>
<td>0</td>
<td>0</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Epistaxis plus other manifestations:</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Easy bruising</td>
<td>17</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Positive family history</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Bleeding after T &amp; A and dental extraction</td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
tility. There is, however, no actual defect in
the coagulation mechanism. In vascular hemophila (affecting seven of the children) an
apparently identical abnormality of the small
vessels is accompanied by a deficiency of anti-
hemophilic globulin, leading to a dual defect.

Both pseudohemophilia and vascular hemophila 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 sug-
gesting 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 PT 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 consump-
tion and thromboplastin generation tests are re-
quied 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 dis-
order. Children with mild to moderate PTC,
PTA and AHC 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 ex-
amination 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 epis-
taxis, of course, occurs as a manifestation of
several systemic diseases such as rheumatic
fever, glomerulonephritis, leukemia, sickle-cell
anemia, and others. As mentioned earlier, how-
ever, 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 experi-
ence epistaxis has rarely been the sole present-
ing manifestation of the child with thrombo-
cytopenic 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 pa-
tients 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 at-
tempt to eliminate a seemingly abnormal blood
vessel. We were not impressed that any or-
ganic 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. Hsiu:** 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.

**Acknowledgment**

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