Recurrent epistaxis in children: When should we suspect coagulopathy?

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Objective
The objective of this study was to evaluate children and adolescents with recurrent epistaxis and to spot variables that may predict underlying bleeding disorder.

Study design
This was a prospective study.

Materials and methods
Thirty-six children with recurrent epistaxis were recruited from ENT outpatient clinic as well as emergency room of Cairo University children hospital. Patients with systemic causes for epistaxis were not included. All patients were subjected to basic workup to differentiate between ENT and hematological causes of epistaxis. Accordingly, extended ENT investigations or coagulation studies were performed.

Results
Results showed 47.6% incidence for bleeding disorders, 39.7% incidence for ENT causes, and 12.7% incidence for idiopathic causes. Reported coagulopathies included idiopathic thrombocytopenic purpura with an incidence of 26.7%, platelet functions disorders such as Glanzmann's thrombasthenia (13.3%) and Bernard–Soulier syndrome (10%), aplastic anemia (3.3%), hemophilia A (3.3%), Von Willebrand disease (3.3%), and unclassified (40%). Reported local ENT causes included bacterial rhinitis (88%) (lodged foreign body was found in 1/3 of these patients, digital trauma by habitual nose picking was detected in another 1/3, and the remaining 1/3 represented cases of complicated previous viral rhinitis), nasal allergy (8%), and adenoid (4%).

Conclusion
The study recommended some statistically significant predictors for bleeding disorders such as positive consanguinity, high epistaxis bleeding score, presence of other bleeding sites, low hemoglobin level and platelet count, and elevated activated partial thromboplastin time.

Keywords:
children, coagulation studies, ENT causes, ENT investigations, hematological causes, recurrent epistaxis, underlying bleeding disorder

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Introduction

Epistaxis or nosebleeds is a common pediatric complaint. Severe and recurrent forms of epistaxis constitute a major trouble to the parents and treating doctors. According to the literature, causes of recurrent epistaxis look different between adults and children regarding frequency distribution of such causes.

In adults, up to 1/3 of the cases are associated with medications such as NSAIDs, warfarin, and aspirin [1,2]. Watkinson [3] stated that epistaxis in adults is often seen with hypertension or arteriosclerosis. Varshney and Saxena [4] in a survey conducted on 88 patients found that epistaxis is essentially a problem of elderly population and that cardiovascular disorders play a considerable role as a causative factor. In children, the most common cause is digital manipulation that leads to vascular fragility in the nasal vestibule. Other less common causes include allergic rhinitis, trauma, infections, anatomic variations, benign and malignant neoplasms, and bleeding disorders [5].

As otolaryngologist is the first on call to manage the nose bleeding (by local drugs, packing, cautery) and to look for local causes for such bleeding, he should be properly oriented with the possibility of the presence of underlying bleeding disorder. The latter group will only respond to replacement therapy by plasma or deficiency factors. Further management of such patients need consultation and follow-up inside hematology clinics. The aim of this study was to study the frequency distribution of different causes of recurrent epistaxis in children and to evaluate variables of significance that may be considered as predictors of coagulopathy in such children.

Materials and methods

This study included 63 children with recurrent epistaxis who were followed up in ENT outpatient clinic or emergency room of Cairo University children hospital through the period from June 2010 to December 2010.

Only those children who were proved to be free from systemic cause (hypertension, liver or kidney failure, rheumatic heart disease, collagen vascular disease, malignancy, under medications contributing to bleeding tendency) were included in this study. The severity of epistaxis was graded using the Katsanis epistaxis scoring system, where epistaxis was considered mild with 0–6 score and severe with 7–10 score (Katsanis *et al.*) [6]. The study was approved by the committee of ethics and research related to the Otolaryngology Head and Neck Surgery Department in Cairo University. All patients had preformed consent signed from their parents for participating in the study.

All patients were subjected to the following:

(1) Detailed history taking with emphasis on the age at onset, recurrence rate, severity, unilateral or bilateral, precipitating factors, other bleeding sites, family history, consanguinity, and treatment received.
(2) Clinical examination for blood pressure, pallor, ecchymosis, lymph nodes, hepatosplenomegaly, and hemarthrosis.
(3) Local ENT examination such as follows:
    (a) Anterior rhinoscopy using nasal speculum to examine the anterior nasal cavity and nasal septum.
    (b) Nasal endoscopy to examine the nose and nasopharynx for inflammatory signs, foreign bodies, polyps, telengectasia, or hemangioma.
    (c) Extended investigations in the presence of local cause such as polyps, hemangioma, mass, or allergy. These included plain radiograph, computed tomography, MRI, nasal discharge culture, blood profile for eosinophils, IgE, and allergic skin tests.
(4) Blood investigations such as:
    (a) complete blood count,
    (b) hemoglobin concentration,
    (c) mean corpuscular volume,
    (d) basic coagulation screen [bleeding and clotting time, prothrombin time, prothrombin concentration, activated partial thromboplastin time (APTT)],
    (e) Extended investigations in patients with abnormal coagulation results. These included platelet functions, clotting factors, and bone marrow aspirate or biopsy.

Statistical analysis

Data were statistically described in terms of mean±SD, median and range, or frequencies and percentages when appropriate. Comparison of numerical variables between the study groups was carried out using the Student *t*-test for independent samples. For comparing categorical data, the χ^2-test was performed. The exact test was used instead when the expected frequency was less than 5. *P* values less than 0.05 was considered statistically significant. All statistical calculations were performed using computer program SPSS (statistical package for the social science) version 16 for Microsoft windows.

Results

This study included 34 (54%) boys and 29 (46%) girls. The patients' age ranged from 2–14 years with a mean of 3.4 years. Twenty-eight (44.4%) patients had positive family history with similar condition. Twenty-four (38.1%) patients had positive consanguinity. Epistaxis was bilateral in 41 (65.1%) patients, whereas it was unilateral in the rest of the patients. Thirty-six (57.1%) patients had mild epistaxis (score 0–6) and 27 (42.9%) patients had severe epistaxis (score 7–10). Thirty (47.6%) of our patients had bleeding symptoms other than epistaxis. Ecchymosis was the most common symptom in 28 (44.4%) patients; oral bleeding was observed in eight (12.7%) patients, hematemesis in six (9.5%) patients, bleeding per rectum in one (1.6%) patient, hematuria in three (4.8%) patients, and menorrhagia in six (9.5%) patients (Figs 1 and 2).

In view of laboratory results, epistaxis was attributed to hematological causes in 30 (47.6%) patients, 18 (60%) of them were diagnosed with bleeding disorder. The diagnosis was as follows: idiopathic thrombocytopenic purpura (ITP) in eight patients, Glanzmann's thrombasthenia in four patients, Bernard–Soulier syndrome in three patients, aplastic anemia in one patient, hemophilia A in one patient, and Von Willebrand disease (VWD) type 1 in one patient. The remaining 12 (40%) patients were considered as having unclassified bleeding disorder, as they showed other bleeding manifestations in addition to the recurrent episodes of epistaxis with a significant family history of bleeding requiring replacement therapy and plasma and blood transfusion with free ENT examination (Fig. 3).

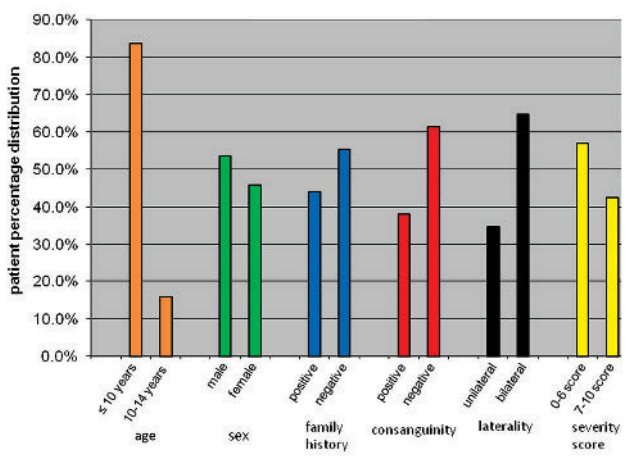
Twenty-five (39.7%) patients had positive local nose causes of epistaxis. Eighty-eight percent of these patients had bacterial rhinitis diagnosed by positive bacterial culture for nasal discharge; the most common organisms included *Streptococcus pneumonia*, *Haemophilus influenzae*, and *Moraxella catarrhalis*. One-third of such patients were found to be habitual nose pickers by history taking and had positive crusting of the anterior nasal septum due to digital trauma. Another 1/3 of the patients showed lodged

nasal foreign body diagnosed by unilateral foul nasal discharge, inflamed mucosa, and granulation tissue around foreign body, and on endoscopic examination foreign body was detected and removed. The remaining 1/3 of the patients most probably represent cases of complicated previous viral rhinitis. Eight percent of the patients had allergic rhinitis; anterior rhinoscopy showed congestion of nasal mucosa and blood-streaked mucus. Extended ENT investigations showed increased IgE, eosinophilia, and positive skin test. One patient was complaining of frequent bilateral nasal obstruction in addition to epistaxis; on examination the child showed adenoid facies, and radiograph of the nasopharynx showed hypertrophied adenoid encroaching on the airway (Fig. 4).

Eight (12.7%) patients were considered idiopathic, as they were ENT free and showed neither bleeding from other sites nor abnormal laboratory results. Hence, hematological causes of epistaxis were more common (47.6%) than the ENT causes (39.7%), whereas 12.7% of patients with epistaxis were considered idiopathic (Fig. 5).

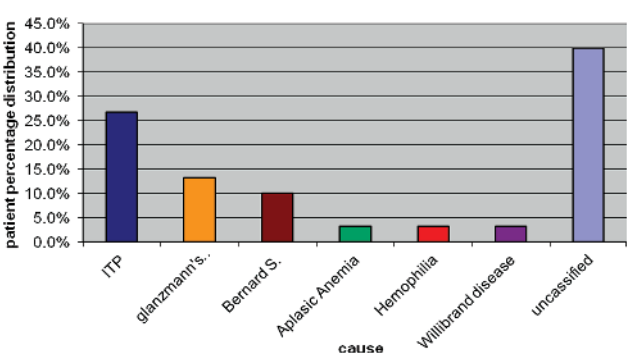
Eighty-one percent of patients were treated with tranexamic acid (kapron, which is an antifibrinolytic drug acting by competitively inhibiting the activation of plasminogen to plasmin). Nasal packing was required in 65.1% of patients; replacement therapy, blood transfusion, platelet and plasma transfusion in 41.3% of patients; and cautery in 9.5% of patients (Fig. 6).

Figure 1



Frequency distribution of different variables related to the total sample ($n = 63$).

Figure 3

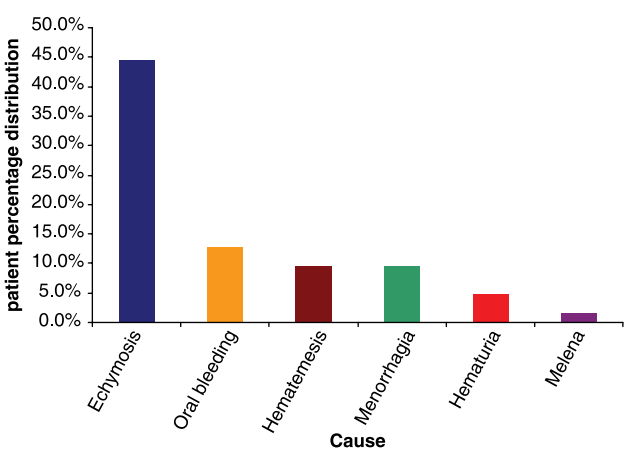


Frequency distribution of hematological patients according to the cause ($n = 30$). ITP, idiopathic thrombocytopenic purpura.

Discussion

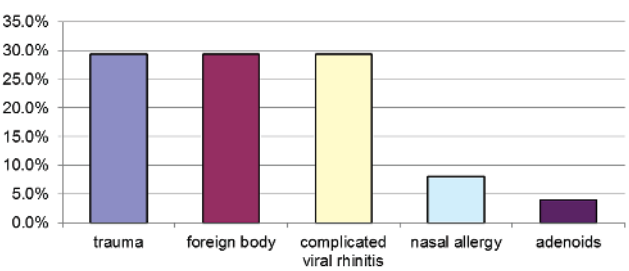
Epistaxis is a common problem among children and also represents a common emergency in otolaryngology [7]. It is crucial in recurrent and severe cases to diagnose the cause of bleeding being local or secondary to inherited systemic coagulopathy. Frequency distribution for causes of epistaxis looks unique in children as compared with adults. Therefore, identification of the cause is important, as it strongly reflects the management plan being totally followed by otolaryngologist or pediatric

Figure 2



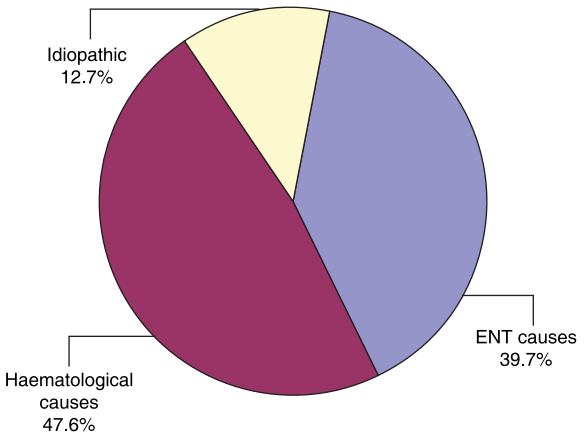
Other bleeding sites in patients with recurrent epistaxsis.

Figure 4



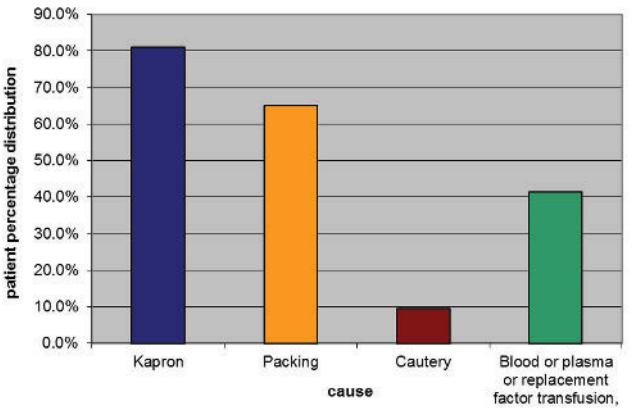
Frequency distribution of ENT patients according to the cause ($n = 25$).

Figure 5



Frequency distribution of the total sample according to ENT, hematological, and idiopathic causes.

Figure 6



Treatment options.

hematologist. In the present study, according to basic ENT examination and coagulation study carried out in all patients and followed by extended ENT investigations or coagulation study indicated for some patients, the results were as follows.

Blood dyscrasias constituted 47.6% of the sample compared with 33, 30, and 10% incidence reported by Sandoval *et al.* [8], Viljoen [9], and Dizdar *et al.* [10]; this reflects the importance of diagnosis of such group of patients as their management plan will be followed in hematology units. Local causes constituted 39.7% incidence; such large sector will be totally managed by otolaryngologist, as it mainly includes trauma, deviated septum, and allergic rhinitis. Idiopathic causes where both ENT examination and coagulation study were free constituted 12.7% compared with 9.2 and 11.2% reported by Viljoen [9] and Hussain *et al.* [7].

In this study, bacterial rhinitis was reported in 88% of patients with local causes for epistaxis. One-third of such patients showed crusting and excoriation of the anterior nasal septum due to digital trauma induced by nose picking, which is a common behavior in children. This coincides with the study by Kubba [11]. *Staphylococcus aureus* seems to have a role in the process; colonization of the nasal cavity by it produces low-grade chronic inflammation leading to septal neovascularization that with digital trauma may explain the sequence of events resulting in recurrent epistaxis [12]. Lodged foreign body was detected in 1/3 of patients presenting with bacterial rhinitis in whom epistaxis was associated with unilateral foul discharge. Foreign bodies that cause epistaxis usually have sharp edges, irritating chemical properties, and/or porosity. Figueiredo *et al.* [13] in their study on 420 patients with nasal foreign body stated that epistaxis is the most common complication of lodged nasal foreign body. Ogunleye and Sogebi [14] in their study on 106 patients with nasal foreign body found that the incidence of epistaxis as a clinical presentation was 5.7%. They also stated that the most common nasal foreign body was seeds (32.1%).

In the present study, nasal allergy was reported in 8% of patients with local causes of epistaxis. There were associated sneezing, itching, and nasal blockage. Anterior rhinoscopy revealed hyperemic mucosa, blood-streaked mucus, and increased vascularity and friability. Extended ENT investigations revealed eosinophilia, increased IgE, and positive skin test; such findings were also reported by Watkinson [3], Ahmad and Zacharek [15], and Deshazo and Swain [16].

The present study showed that adenoid constituted an incidence of 4%. Adenoid per say was not reported in the literature as a common cause of epistaxis; however, nasal obstruction leads to a turbulence of air flow anterior to this obstruction that has a drying effect increasing the opportunity for mucosal disruption and epistaxis.

Rank distribution for different inherited coagulopathies in this study shows: ITP (26.7%), Glanzmann's thrombasthenia (13.3%), Bernard–Soulier (10%), aplastic anemia (3.3%), hemophilia A (3.3%), VWD type 1 (3.3%), and unclassified bleeding tendency (40%). Sandoval *et al.* [8] in their study in 2002 including 59 children with recurrent epistaxis reported VWD type 1 to have the highest rank (33/59) followed by platelet aggregation defect (10/59), ITP (7/59), Bernard–Soulier (2/59), factor VIII deficiency (3/59), factor VII, IX, XI deficiency in one child each (3/59), and presence of coagulation inhibitor (1/59). Chandra *et al.* [17] reported ITP

with 13.4% incidence. Sidonia *et al.* [18] in their study including 298 patients reported low Von Willebrand factor (VWF) in 34% of patients, platelet aggregation defect in 16%, VWD in 8%, and normal hemostatic test in 42% of patients. Mokhtar *et al.* [19] in over 16-year study in Egypt reported Glanzmann's thrombasthenia in 11.2% of patients and VWD in 6.6% of patients. Unclassified coagulopathy in this study had an incidence of 12/30 (40%); they showed abnormal APTT and extended investigations did not show classified error. El-bostany *et al.* [20] reported 30% of unclassified coagulopathy.

With respect to the predictors of bleeding disorder, increased bleeding score was considered statistically significant predictor in this study ($P = 0.001$). The same finding was confirmed by Tosetto *et al.* [21] and Sidonio *et al.* [18] who reported that higher epistaxis bleeding score increases the likelihood of an underlying bleeding disorder. The presence of other bleeding sites also proved statistically significant in predicting inherited coagulopathy in this study ($P = 0.001$). Menorrhagia in a study on 106 female adolescents with recurrent epistaxis proved to show 17% incidence among patients proved to have inherited coagulopathy [22]. Further studies reported menorrhagia to be commonly present with epistaxis in Glanzmann's thrombasthenia and Bernard–Soulier [23]. Positive consanguinity was statistically significant predictor of blood disease in this study ($P = 0.004$), which was in agreement with the study by Mokhtar *et al.* [19] and Elden *et al.* [24]. Shaw *et al.* [25] confirmed the previous finding with a positive predictor value of 45%. In contrast, Sidonia *et al.* [18] reported neither consanguinity nor the presence of other bleeding sites as predictors of VWD, low VWF, or platelet aggregation defect. A trend was found in those patients with epistaxis at a younger age [24]. Anemia with low hemoglobin as well as low platelet count showed statistical significance in this study ($P = 0.001$) as also reported by Damrose and Maddalozzo [26]. APTT proved to be a significant predictor in this study ($P = 0.001$), which was in agreement with the study by Sandoval [8] and Gerlinger *et al.* [27]; however, the value of prothrombin time (PT) and APTT as a predictor was doubted by Shaw *et al.* [25]. Table 1 shows comparison of all clinical variables of the patient group with an underlying hematological cause as opposed to that of a nonhematological cause.

Table 1 Comparison of all clinical variables of the patient group with an underlying hematological cause as opposed to that of a nonhematological cause

Variables	Hematological ($n = 30$)	Nonhematological ($n = 33$)	*P*-value
Age (mean) (years)	6.6	5.6	0.294 (NS)
Sex [*n* (%)]			
Male	15 (50)	19 (57.6)	0.547 (NS)
Female	15 (50)	14 (42.4)	
Consanguinity [*n* (%)]			
Positive	17 (57.6)	7 (21.2)	0.004 (S)
Negative	13 (42.4)	26 (78.8)	
Family history [*n* (%)]			
Positive	14 (46.7)	14 (42.4)	0.735 (NS)
Negative	16 (53.3)	19 (57.6)	
Epistaxis score [*n* (%)]			
Mild	6 (20)	30 (90.9)	<0.001 (S)
Severe	24 (80)	3 (9.1)	
Other bleeding symptoms [*n* (%)]			
Positive	30 (100%)	0	<0.001 (S)
Negative	0	33 (100)	
Laboratory tests (median)			
Hb (g/dl)	10.1	12.0	<0.001 (S)
Plt count (×10/l)	199	364	<0.001 (S)
APTT (s)	30.0	27.0	<0.001 (S)

APTT, activated partial thromboplastin time; Hb, hemoglobin; Plt, platelet; S, significant.

Conclusion

Causes of recurrent epistaxis in children in this study show unique frequency distribution compared with that of adults reported in the literature review. Underlying inherited bleeding tendency represents a major sector. Therefore, recurrent epistaxis may be a leading sign of coagulopathy. Suggested predictors of underlying bleeding disease in this study include high epistaxis bleeding score, presence of other bleeding sites, positive consanguinity, low hemoglobin level, low platelet count, and prolonged APTT. Basic suggested workup in this study that initiates with good history, local ENT examination, and basic coagulation study (bleeding time, PT, PC, APTT) is effective in differentiating between hematological and local causes. Extended workup is needed after differentiation, including advanced imaging modalities and endoscopy, in local causes and advanced coagulation studies are required in hematological causes.

Acknowledgements

Conflicts of interest

None declared.

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