Challenges in the Management of Bleeding Disorders in Nigeria

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INTRODUCTION

Bleeding disorders (BDs) are characterized by prolonged or abnormal bleeding due to disorders of blood vessels, platelets, or coagulation factors. They are sometimes referred to as clotting abnormalities or coagulopathies by different practitioners. The prevalence of BD is variable depending on the population being studied, while the prevalence is about 29%–47% in women presenting with menorrhagia.[1,2] von Willebrand disease (VWD) is thought to be the most common inherited BD affecting 1% of the general population.[3] Acquired BDs are more common than inherited BDs and are usually secondary to multiple coagulation defects.[4] These disorders could be inherited or acquired. BDs are also classified based on the mechanism of defect.

The inherited BDs are grouped into disorders due to coagulation factor deficiency, platelet and fibrinolytic disorders, and vascular and connective tissue disorders. The most common inherited coagulation disorders include hemophilia A (factor VIII deficiency), hemophilia B (factor IX deficiency), factor XI deficiency, and VWD.[5,6] Acquired causes of BD include those resulting from thrombocytopenia (which could be immune mediated, drug induced, or due to hematological malignancies) or disseminated intravascular coagulation (DIC), liver and renal diseases, acquired antibodies against clotting factors, and Vitamin K deficiency, among others.

Patients with BD can present with a myriad of symptoms ranging from mucocutaneous bleeding as

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seen in platelet disorders such as thrombocytopenia or VWD to haemorrhoses, hematuria, intramuscular, intracerebral, and retroperitoneal hemorrhages seen in severe hemophlias A and B, severe VWD, and severe deficiency of factors VII, X, and XIII. Spontaneous bleeding, postcircumcision bleeding, and injury-related bleeding can also occur.[11] In addition, menorrhagia could be the first and only symptom of a patient with BD. Given the spectrum of presentation, the different departments or specialties could be the portal of entry for these patients. Furthermore, some may have presented for other medical or surgical problems and the BD detected during examination and laboratory investigations.

A detailed history taking and physical examination as well as basic and readily available investigations is essential in making a diagnosis of BDs. Management of BDs is laden with numerous challenges, which include lack of adequate health-care facilities, low awareness among medical practitioners, lack of multidisciplinary team approach, insufficient number of specialists and specialist centers, lack of effective medical insurance, lack of factor concentrates as seen in hemophiliaas, insufficient government funding, disease underregistration, and lack of availability of treatment products.[7-9] A study carried out in northern part of Nigeria stated that one of the problems facing the Nigerian health system is limited access to health facilities and that most hospitals in Nigeria are privately owned by individuals and religious organizations which may lack adequate physical structures and equipment, adequate and skilled workforce, and service delivery in general.[10] Reding and Cooper[11] pointed out that barriers to effective diagnosis and management of a BD are due to lapses in knowledge and practice which could be improved by more effective education.

This study aims to determine the capacity of doctors working in hematology units to handle BDs in Nigerian health institutions and also to determine the degree of availability of infrastructure, equipment, and treatment options required for its management.

**Materials and Methods**

This was a descriptive study conducted in Nigeria during the annual scientific conference of the Nigerian Society for Haematology and Blood Transfusion which held in October 2016.

A structured questionnaire was distributed to hematologists who were in attendance at the conference and practicing in Nigerian institutions involved in the management of BDs. Information including name of institution, number of hematologists present, the availability and functionality of infrastructure and equipment, available investigations, and treatment options necessary in handling BD were sought for. On completion, these questionnaires were retrieved and data were analyzed using the Statistical Package for the Social Sciences (SPSS) software Version 21 (IBM, Chicago, Illinois, USA). Results were presented in tables and as frequencies and percentages.

**Results**

From the 72 hematologists who attended the meeting, 55 responded giving a response rate of 76.4%. These 55 hematologists were from 27 Nigerian health institutions. Hematologists were involved in patients’ management in the clinical departments of all institutions; however, hematologists in only 21 (77.8%) institutions consulted pediatric patients [Table 1].

The most common laboratory section available for work was the general hematology laboratory present in 25 (92.6%) out of the 27 centers. This was closely followed by the coagulation laboratory and blood bank available in 88.9% of the centers. The least available laboratory sections were the research laboratory (51.9%) followed by the residents’ laboratories (59.3%), as shown in Table 1.

Hemoglobin (Hb) concentration and packed cell volume were the most frequently carried out investigations (100%). This was closely followed by the full blood count (including platelet count), both equally carried out in 26 (96.3%) centers. Prothrombin time (PT)/international normalized ratio (INR) and activated partial thromboplastin time (APTT) were the coagulation tests frequently carried out in twenty (77%) institutions.

**Table 1: Coverage of services and laboratory space**

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<th>Total n (%)</th>
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<td>Wards</td>
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<td>Pediatrics</td>
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<td>Obstetrics and gynecology</td>
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<td>Accident and emergency</td>
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<td>Research laboratory</td>
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<td>Residents’ laboratory</td>
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Most of the other coagulation investigations were not carried out in those centers. Majority of the institutions did not carry out Hess/Rumple–Leede test (76.9%), inhibitor assay (75%), fibrinogen assay (60%), factor assay (58.3%), and mixing studies (47.8%). von Willebrand factor assay was only sparingly done in two (8.4%) centers. DNA-based tests were never carried out in 22 (88%) institutions as summarized in Table 2.

Of the available equipment, microhematocrit centrifuge was the most used (100%) followed by the full blood count auto-analyzer, 24 (88.9%). None of the respondents had thromboelastography or rotational thromboelastometry in their centers, while most centers did not have multiplate (84.2%), polymerase chain reaction (PCR) machine (71.4%), apheresis machine (50%), coagulometer (47.8%), and cold centrifuge (43.4%). The PCR, apheresis, and ELISA machines were present but had never been in use in 19.1%, 12.5%, and 8.3% of the institutions, respectively [Table 3].
Challenges in managing bleeding disorders

Platelet concentrates are...
encountered BDs in clinical practice are the acquired BDs, with DIC being one of the most common types. Bleeding is the most readily apparent clinical feature of DIC. These may explain why respondents reported to have encountered DIC most in their practice and being one of the most common acquired BDs. As a result, whole blood (in the absence of component therapy) and tranexamic acid were seen as the most commonly available treatment options in this study. At the same time, 88% of respondents accepted to frequently use steroids, the first-line treatment for ITP, which is equally one of the common acquired BDs.

This study also shows that less than half of the participants had had a formal training in the field of hemostasis in the previous year, thereby impacting negatively on the care of patients with BDs. Although continuous medical education (CME) is required by the Nigerian Medical Association before annual renewal of doctors’ practicing license, it does not specify the specialty areas for doctors. Therefore, hematologists may have received CMEs in some other aspect of hematology, excluding the field of hemostasis.

The establishment of basic minimum tests, equipment, treatment options, and personnel training for effective management of patients with BDs in these institutions is necessary. These include tests such as full blood count, bleeding time, PT/INR, APTT, thrombin time and fibrinogen assay, equipment to produce blood components, and other treatment options such as the factor concentrates.

### Conclusion

There is significant deficiency in most centers in Nigeria to adequately diagnose the underlying cause in a bleeding patient. Therapeutic options are also limited to whole-blood transfusion and tranexamic acid as there is poor access to component therapy or clotting factor concentrates. This study reveals some of the challenges in the care of patients with BDs in a resource-poor setting.

### Recommendations

Massive funding is required for major Nigerian health institutions in order to provide infrastructure, facility, and training required in the diagnosis and treatment of patients who present with BDs.

### Financial support and sponsorship

Nil.

### Conflicts of interest

There are no conflicts of interest.

### REFERENCES