Comprehensive Spectrum of Inherited Bleeding Disorders from South West Iran

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AIM

The aim of the present study is to assess the frequency, magnitude and diversity of different bleeding disorders in south west Iran.

MATERIALS AND METHODS

A total of 415 cases with inherited bleeding disorders were assessed from April 2010 to July 2011. A comprehensive evaluation such as diversity presentations, clinical manifestations, and demographic data were recorded. This is a descriptive cross sectional study that was conducted at Ahvaz Jundishapur University of Medical Sciences in Khuzestan province. Data were collected through a questionnaire form and statistical analysis was done by using Statistical Package for the Social Sciences version 17.

RESULTS

A total of 415 patients were diagnosed at Shafa hospital. The results can be divided and sub-divided in two main categories as shown in Figure 1. The following points were also observed;

- Amongst patients, 189 cases (45%) had no history of family involvement and the rest of them (55%) reported a positive family history in their close relatives.
- Mouth and nose bleeding were the first presenting symptoms in Glanzmann and VWD.
- Joint, post circumcision, mouth and nose bleeding were the initial symptoms in coagulation disorders.
- Surprisingly hemarthrosis was the first symptom in 9 patients (17%) of Glanzman's syndrome and 6 patients (10.5%) of VWD.

The severity of disease among 195 patients of hemophilia and 39 patients of hemophilia B were shown in Figure 2. The following observations were also made:

- 6 patients (3%) had inhibitor in hemophilia A group (two, low titter and four, high titter).
- All inhibitors were in severe patients.
- None of hemophilia B and VWD patients had inhibitor.
- 50% of hemophilia A, 66.7% of hemophilia B and 29.3% of VWD had target joint.
- 8 (4.1%) of hemophilia A and 2 (5.1%) of hemophilia B patients were female.
- Routine viral screening in all patients was as follows: Positive HBS Ag: 0%, Positive HBS Ab: 87.6%, Positive HCV Ab: 8.2 %(confirmed by PCR) and only 1 patient had HIV positive.

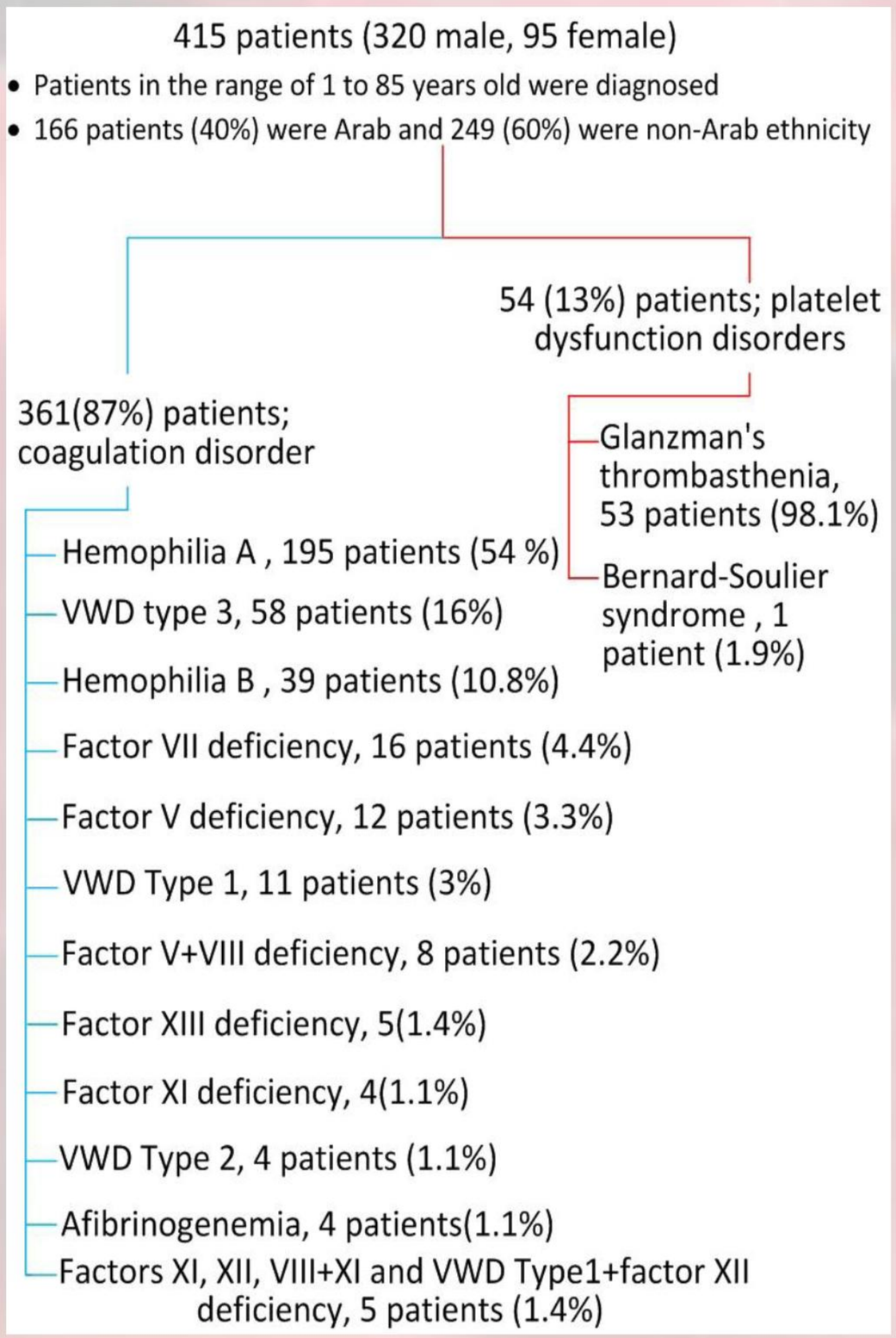

Figure 1: Results of 415 patients diagnosed in south west Iran

CONCLUSIONS

In spite of their apparent rarity, Iran and Khuzestan province have a considerable number of patients with inherited bleeding disorders. A large number of these patients are belonged to hemophilia. In south west Iran the number of VWD and Glanzman's syndrome is considerable. However, these diseases have not a great role in national burden of diseases but, because of high morbidity and treatment expenditures the knowing of all dimensions of these diseases may provide a clue for the health sector providers and experts in this field.

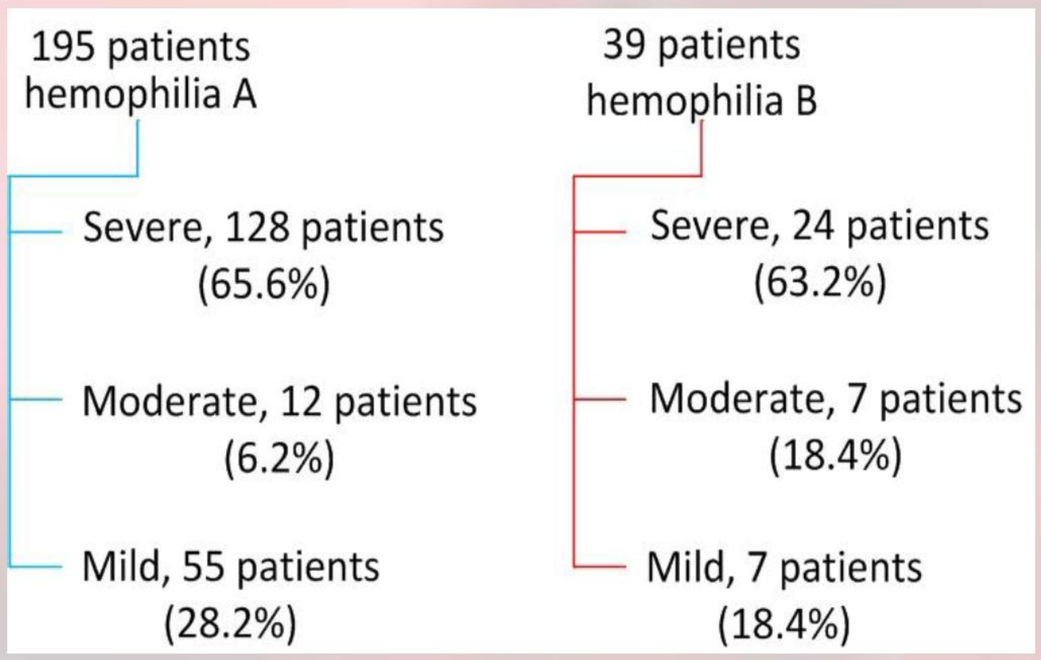

Figure 2: The severity of disease among hemophilia A & B patients

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