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# Epidemiology of hemophilia at the Hospital of Rabat:About 263 cases

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## ABSTRACT

*The haemophilia is a constitutional hemorrhagic disease of recessive transmission linked to X chromosome. The objective of this work is to index the different cases in our service. Material and methods: During 45 months (April 2008 – December 2012), we account 263 haemophiliac cases diagnosed in our laboratory. Results: Our results interest 263 haemophiliac cases, which means 198 cases of haemophilia A (75,3%) and 65 cases of haemophilia B (24,7%) diagnosed. 172 patients have an age less than 15 years old with a median of age as 13 years (limits 9 months - 39 years). The annual incidence of haemophilia is 70 cases. 132 persons had clinical symptomatology whereas the others were asymptomatic. These symptoms were arthropaty (62,14%), joint bleed (23,48%), haematoma (5,30%), nosebleeds, bleeding gums (4,54%) and ecchymosis (4,54%).According to the type of haemophilia: The severe forms of haemophilia (FVIII or FIX ≤ 1%) are 57,08% (115/198) for haemophilia A and 64,63% (47/65) for haemophilia B. Discussion: The diagnosis and classification of the haemophilia are possible in routine. Through the data of literature we propose a diagnostic strategy which makes it possible to consolidate the analysis of the constitutional haemostasis hemorrhagic diseases in particular of the haemophilia. Conclusion: The assumption of responsibility of the haemophilia starts with this stage. Our hope is to be able to extrapolate this study at the national level to set up a national file.*

**Keywords :** Haemophilia, Biological diagnosis.

## INTRODUCTION

Hemophilia is a bleeding disorder inherited as constitutional a recessive X-linked, due to a deficiency of factor VIII (hemophilia A) or factor IX (hemophilia B) of coagulation. This condition is the most common serious constitutional hemorrhagic diseases [1-3]. Its incidence is 1/5000 newborn males in hemophilia A and hemophilia B in 1/30000 [1-3]. The objective of this work is to identify cases of hemophilia diagnosed in our laboratory.

## MATERIALS AND METHOD

This is a retrospective study which focused on a series of 263 cases of hemophilia diagnosed during a 45-month period (April 2008 - December 2012). The study and use of the data were based on the results complemented by hemostasis and archived in the service record. The review carried out on the semi-automatic analyzer ST art, included:

- Prothrombin Time (NeoplastinCl 10, Stago)

- Partial thromboplastin time / Activator (PTTA 5, Stago)

- Determination of FVIIIc activity (STA deficient VIII, Stago)

- Determination of FIXc activity (STA deficient IX, Stago).

Depending on the size of the deficit in activity in haemophilia [4], have been distinguished:

- Severe forms FVIII or FIX levels ≤ 1%.

- Moderate forms: FVIII or FIX between 1 and 5%.

- Minor forms: FVIII or FIX between 5 and 30%.

## RESULT AND DISCUSSION

Our results concerned 263 cases of hemophilia over a period of 45 months with an incidence of 70 cases per year. There were 198 cases of hemophilia A and hemophilia B 65 diagnosed cases (Figure 1). The median age was 13 years (range 9 months - 39 years). 172 patients were under 15 years, a proportion of 65.39% (Figure 2).

132 patients (50.2% of patients) had a license while others were asymptomatic clinical symptomatology.

Various clinical signs were collected (Table I).

The distribution of cases of hemophilia A and hemophilia B is made according to their severity namely major forms, moderate forms and minor forms (Figure 3).

A major diagnostic difficulties constitutional bleeding disorders of hemostasis is the fact that many of them are asymptomatic. The diagnosis of hemophilia is based partly on a careful history looking for a personal history of bleeding and / or family as haemarthrosis or bleeding during circumcision, dental extractions, tonsillectomies and 'adenoidectomy; and secondly, on the haemostasis balance that allows an isolated before elongation aPTT activator (TCA), characterization of FVIII deficiency or FIX and for the presence of an inhibitor [1-5]. Bleeding time, platelet count, prothrombin time, thrombin time and fibrinogen are normal. The dosage or FVIIIcFIXc is usually done by a chronometric method in one time with a deficient plasma or FVIII FIX. This assay allows to determine the severity and long-term prognosis of the disease. It has recently been reported in rare cases where moderate FVIII assay technique at a time was set default deficits, it is useful in this case to use a chromogenic technique. Furthermore, it is essential to complete the phenotype of hemophilia A by assaying vWF antigen in the conductive [2, 6]. Finally, the antigen assay may be carried out during the diagnosis to differentiate hemophiliacs CRM (Cross Reacting Material) characterized by positive presence of CRM and antigen negative (no antigen) [7,8].
In our series, diagnosis and classification of hemophilia are available routinely. The majority of patients with haemophilia A (75.3%), these results are consistent with those in the literature which states that hemophilia A is four times more common than hemophilia B [8]. Furthermore, H. Langar et al [9] showed that 87.1% of patients with hemophilia A and hemophilia B. 12.9% Also, our study showed that the severe forms, moderate and minor (hemophilia A and B combined) accounted for 58.95%, 29.27% and 11.78% respectively. H. Langar et al showed that 61.5% of patients had severe hemophilia and 31.4% moderate hemophilia.

Severe hemophilia was predominant for all age groups, particularly for patients aged between 1 and 2 years where the percentage reached 87.09%, this can be explained by the early onset of symptoms of this disease and consequently how early diagnosis.

Our diagnostic strategy must be supported by antigenic study. The adherence of patients' families must be consolidated in order to detect potential conductive and boys could be achieved.

## CONCLUSION

The diagnostic approach constitutional bleeding disorders of hemostasis must be rational combining both clinical and laboratory data. We were interested to hemophilia, which can be a very debilitating disease if its management is not early. The true prevalence of hemophilia in Morocco is not known where the interest of a national registry.

**Table I: Distribution of hemophiliacs effective symptomatically:**

| Symptom | Number of cases | Percentage (%) |
|---|---|---|
| Arthropathy | ***82*** | 62,14% |
| Hemarthrosis | **31** | 23,48% |
| Hematoma | **7** | 5,30% |
| Epistaxisand gingival | **6** | 4,54% |
| Bruise | **6** | 3,54% |
| Grand total | 132 | 100% |

**Figure 1: Distribution of patients by type of hemophilia.**

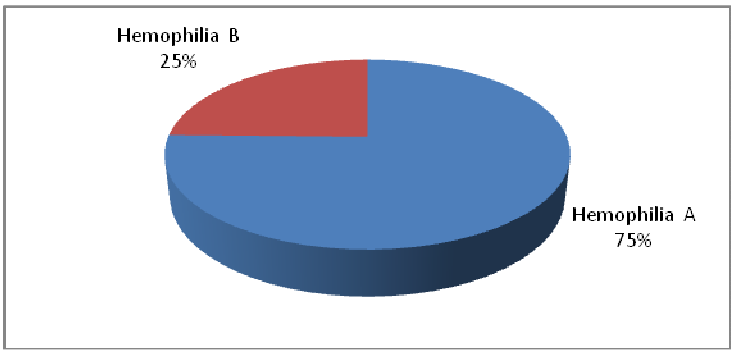



**Figure 2: Distribution of cases of hemophilia according to age group.**

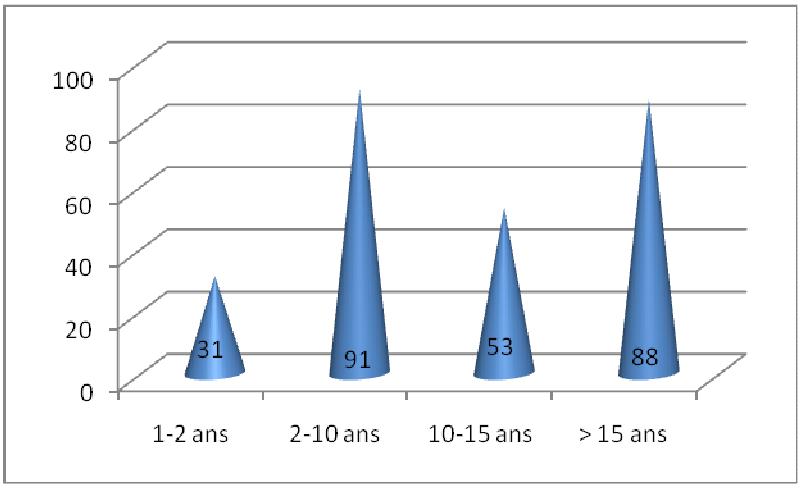
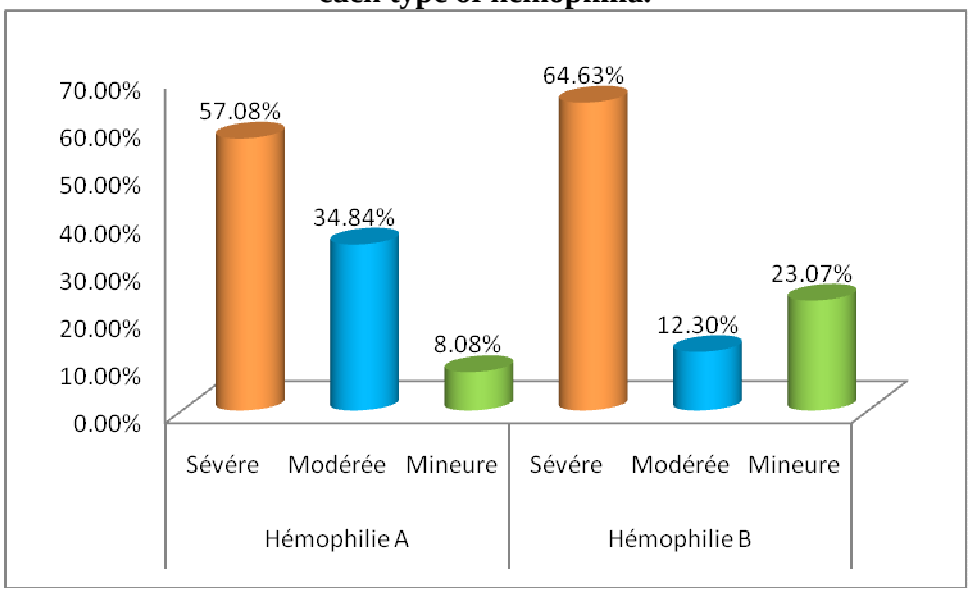



**Figure 3: Distribution by percentage according to the importance of the factor deficiency for each type of hemophilia.**

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