WFH 2016 – Hemophilia-clinical
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Clinical and biological aspects of haemophilic patients in Ouagadougou (Burkina Faso)

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Introduction

Considering the incidence of haemophilia, 1500 haemophilic patients could be diagnosed in Burkina Faso. Because of the lack of medical knowledge on haemophilia and because of the lack of biological diagnosis (when haemophilia was suspected) no case of haemophilia had been registered until April 2015. Thanks to AFATH (Alliance Franco Africaine pour le Traitement de l'Hémophilie) program, the biological diagnosis of Haemophilia is now available in Ouagadougou

Objective of the study

To give a picture of the cohort of haemophilic patients in 2016

Materials and Methods

In 34 patients, haemophilia had been suspected on haemorrhagic manifestations and on the correction of prolonged aPTT when patient plasma was mixed with control plasma.

Factors VIII and IX were measured using a chronometric one-stage aPTT-based assay [1] on a Start 4 (reagents and instrument kindly provided by Diagnostica Stago, France)

We also looked for the presence of anti VIII or IX inhibitor, anti HIV, HCV and HBV antibodies.

Conclusion

The results of this study reflect the emergency to enlarge haemophilia diagnosis and to make haemophilic treatment available.

We have to focus our efforts on information to patients and physicians and on family surveys to increase the detection of the disease.

Thanks to the help of the World Federation of Hemophilia (with the gift of Pharmaceutical Industry) specific antihaemophilic on-demand treatment is now available in Burkina.

A low dose prophylactic treatment should be considered for these young patients.

References

1- « Lab Manual 2010 - World Federation of Hemophilia » - chapter 23, 64-69. http://www.wfh.org
2- Löfqvist, T, I M Nilsson, E Berntorp, et H Pettersson. « Haemophilia prophylaxis in young patients a long-term follow-up ». *Journal of internal medicine* 241, nº 5 (mai 1997): 395-400.

Results

- ***Demographic***

  Over a period of 7 months, 29 severe haemophilic patients (21 HA; 8 HB) were diagnosed

  Median age : 7 (range: 0,7 - 28 years)

- ***Circumstances of diagnosis***

  Circumcision was the main (41 % of cases) circumstance of the first haemorragia

  39% admitted in hospital for haemarthrosis prior to diagnosis

- ***Orthopedic joint score***

  - The orthopaedic joint score (Löfqvist, 1997) evaluated in 20 patients was rather high considering the young age of the patients (table 1)
  - Knees were the target joints in 55 % of cases while ankles were curiously free of haemarthrosis (except for 1 patient)
  - Haematomas were mostly located on the arms (36%)

- ***Family surveys began***

  A family tree of one of them is provided (fig 1)
  showing that haemophilia is a deadly disease in Burkina
  Conductive women remain to be diagnosed

- ***Substitutive treatment***

  Only 38% received a substitutive treatment
  Mostly Freeze Frozen Plasma

- ***Anti VIII / anti IX inhibitors :*** **none**

- ***Status of infectious diseases***

Antibody performed :	**HIV**	**HCV**	**HBV**
Positive results, %	0	3	10

Photo 1: Consequences of multiple untreated hemarthrosis

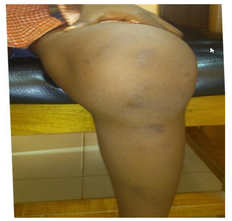
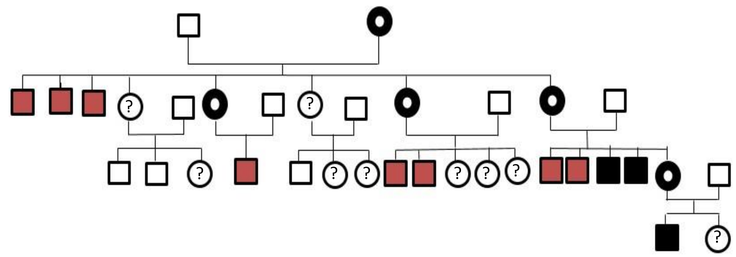


Table 1: Orthopaedic joint score performed in 20 haemophilic patients

Orthopaedic joint score	**Haemophilia A n= 13**	**Haemophilia B n= 7**
[0 – 5[	9	*4*
[5 – 10[	3	1
[10 – 15[	1	1
[15 - 20]	0	1

*Fig 1: **Family tree** from one haemophilic Burkinabe family*



Healthy man □, alive haemophilic patient ■, dead haemophilic patient ■, healthy women but conductive status unknown ?, conductive women ⊙