

# **GUIDELINES FOR THE MANAGEMENT OF HEMOPHILIA IN EGYPT**

**Developed by an Egyptian Expert Panel**

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# BACKGROUND

These guidelines have been developed by an expert panel of hemophilia treaters to support the appropriate management of people with hemophilia in Egypt. Although the guidelines are based primarily on the World Federation of Hemophilia (WFH) Guidelines for the Management of Hemophilia,<sup>1</sup> they aim to address unmet needs and local requirements in Egyptian settings.

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# INTRODUCTION

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The healthcare system in Egypt is quite complex with a large number of public entities involved in management, financing and the provision of care.<sup>1</sup> The World Federation of Hemophilia (WFH) Guidelines for the Management of Hemophilia are widely recognized as a gold-standard document to guide and inform about comprehensive management of hemophilia care.<sup>2</sup> The guidelines here proposed have been developed to address important issues related to the comprehensive management of hemophilia in Egypt, including circumcision and hepatitis C virus (HCV) management. It is important to note that the guidelines should be used as a supplement to WFH recommendations.

# I. GENERAL CARE AND MANAGEMENT OF HEMOPHILIA

## I.1 WHAT IS HEMOPHILIA?

Hemophilia is an X-linked congenital bleeding disorder, caused by deficiency of coagulation factor VIII (hemophilia A) or factor IX (hemophilia B). Hemophilia has an estimated frequency of approximately one in 10,000 births.

Hemophilia A is more common than hemophilia B, representing 80–85% of the total hemophilia population. According to the WFH Global Survey, there were an estimated 5,050 patients with hemophilia in Egypt in 2013.<sup>2</sup>

The characteristic phenotype in hemophilia is the bleeding tendency. Its severity is generally correlated with the clotting factor level (Table I). Hemophilia

should be suspected in patients presenting with excessive bleeding following minor surgery, such as circumcision. Definitive diagnosis should be confirmed through factor assay to demonstrate deficiency of FVIII / FIX.

While bleeding tendency is life-long, people with hemophilia may not present with bleeding symptoms until later in life, for example when they begin walking / running. Management of hemophilia is multifactorial and includes prevention and treatment of bleeding, prevention of long-term joint damage, physical therapy, dental care, vaccinations, nutritional support, genetic counseling and psychological support.

**TABLE I: Relationship of bleeding severity to clotting factor level<sup>1</sup>**

Severity	Clotting factor level	Bleeding episodes
<b>Severe</b>	<1 IU/dl (<0.01 IU/ml) or <1% of normal	Spontaneous bleeding into joints or muscles, predominantly in the absence of identifiable hemostatic challenge
<b>Moderate</b>	1–5 IU/dl (0.01–0.05 IU/ml) or 1–5% of normal	Occasional spontaneous bleeding; prolonged bleeding with minor trauma or surgery
<b>Mild</b>	5–40 IU/dl (0.05–0.40 IU/ml) or 5– <40% of normal	Severe bleeding with major trauma or surgery. Spontaneous bleeding is rare

## I.2 PRINCIPLES OF CARE

The primary aim of care is to prevent and treat bleeding with the deficient clotting factor. Whenever possible, factor deficiency should be treated with factor concentrate (see section 4.2 Product selection). For optimal care, patients with hemophilia should be treated within a comprehensive care setting. A patient's core management team should include a hematologist, nurse coordinator, laboratory specialist, musculoskeletal specialist, physical therapist, orthopedic surgeon, hepatologist and psychological expert. The patient should also have access to a chronic pain specialist, dentist, geneticist, hepatologist, infectious disease

specialist, immunologist, gynecologist / obstetrician and vocational counselor if possible.

In Egypt, children with hemophilia have received minimal dental intervention and their needs required thorough assessment. Oral health education, plaque control, and access to oral care is paramount to improve the oral health of these children.<sup>3</sup>

# I. GENERAL CARE AND MANAGEMENT OF HEMOPHILIA

## I.3 NUTRITION AND VACCINATION

It is important for patients with hemophilia to maintain good nutrition, particularly the absorption of key nutrients such as calcium, iron, vitamins C and D. Management of patients who are overweight or obese is challenging and costly, requiring a constant follow-up on the patient's nutrition, lifestyle, weight and physical activity levels.

Immunization against hepatitis A and hepatitis B viruses is mandatory for all people with hemophilia – including routine hepatitis A vaccination in all patients >1 year of age.<sup>1</sup> Patients >5 years of age should be screened for hepatitis B virus and revaccinated if negative. Vaccination should be subcutaneous, rather than intramuscular or intradermal.

Adequate weight management should be encouraged.

### RECOMMENDATIONS

Appropriate healthcare should be provided to all patients with hemophilia, including nutritional advice and follow-up – calcium, iron (guided by iron level) and vitamins C and D. Routine hepatitis A vaccination in all hemophilic patients >1 year of age is mandatory. Hepatitis B screening is essential for patients with hemophilia >5 years of age, and it is important to revaccinate those who are negative.

## I.4 HOME THERAPY

Early treatment of bleeds minimizes the amount of blood in affected joints, resulting in improved short- and long-term outcomes, such as reduced pain and swelling, as well as reduced joint damage. Home therapy allows a patient immediate access to clotting factor – meaning optimal early treatment is possible – stopping bleeding and reducing long-term damage. Besides the increase of factor level with clotting factor concentrates (or desmopressin in mild hemophilia A) management of muscle and joint bleeds can be achieved through first aid measures such as PRICE: splints (**P**rotection), **R**est, apply **I**ce in the bleeding location, **C**ompression, **E**valuation. In case of mucosal bleeds or dental extractions, antifibrinolytic drugs (i.e. tranexamic acid, epsilon aminocaproic acid) can be an effective adjunctive

treatment. Home therapy is necessary for effective use of prophylactic treatment.

Where home therapy programs are available, there is a reduction in clinic visits and hospital admissions, easing the burden on public health facilities. Nevertheless, it is important to ensure a regular clinic visit (every 3 months) for follow-up. Training and education is very important for both parents and patients, particularly:

- How to self-infuse
- How to keep bleeding records
- How to transport, store and use factor
- How to dispose of needles
- How to handle blood spills
- How to manage complications of therapy

### RECOMMENDATIONS

Home therapy should be encouraged, however, it is important that regular clinic visits continue to ensure good management of hemophilia. First aid measures and antifibrinolytic agents are encouraged as adjuvant therapy. Parents and patients should be educated on all the steps underlying home therapy to build confidence, ensure safety and promote home therapy as an option for the management of hemophilia.

# I. GENERAL CARE AND MANAGEMENT OF HEMOPHILIA

## I.5 PHYSIOTHERAPY

Physiotherapy is important in the management of hemophilia to prevent and treat musculoskeletal damage.<sup>4</sup> In addition to clotting factor concentrates, usually prescribed by the hematologist, management of acute hemarthrosis and chronic arthropathy requires close collaboration between the orthopedic surgeon and physiotherapist. This collaboration, comprising a coagulation and musculoskeletal specialist, is key to effectively prevent hemarthrosis, manage acute joint bleeding episodes, assess joint function and actively treat

chronic arthropathy.<sup>5</sup> Physiotherapy should be initiated as soon as the pain subsides and should be continued to gradually restore full muscle length, strength and function.

The evaluation of joint status, its progress and the impact on quality-of-life represent an integral part of the consultation. A short and easy scoring system can be used to support the evaluation of joint health and should be adopted to monitor patient outcomes.<sup>5</sup>

### RECOMMENDATIONS

It is recommended that the hemophilia treater talks directly with the local physiotherapist to provide education on the special needs of treating someone with hemophilia, specifically regarding the use of factor replacement therapy prior to the physiotherapy session. Physiotherapy maneuvers are best carried out immediately after factor infusion. A short and easy scoring system, such as the Hemophilia Joint Health Score (HJHS) should be adopted to evaluate joint health.

## 2. SPECIAL MANAGEMENT ISSUES

### 2.1 HEPATITIS C VIRUS IN EGYPT

Egypt has the highest prevalence of hepatitis C virus (HCV) in the world, with 14.7% of people aged 15–59 years old affected.<sup>6</sup> The high prevalence is thought to have resulted from transmission during parenteral antischistosomal therapy (PAT) mass-treatment.<sup>7</sup> Current risk of transmission through healthcare and through the community settings is still considered to be substantial.<sup>7</sup>

Knowledge about the viral load distributions in different stages of HCV infection is essential to compare the efficacy of serologic screening and nucleic acid testing in preventing transfusion transmission risk. Studies have been conducted on the HCV-RNA levels in Egyptian blood donors in the pre-seroconversion window period and in later anti-HCV-positive stages of infection.<sup>8</sup>

#### RECOMMENDATIONS

Prevention of HCV transmission in people with hemophilia is a priority of care in Egypt and regular screening is important, even in asymptomatic patients. Patients should be educated on the potential risks underlying the use of blood products and stricter policies are needed to protect them. For HCV-positive patients with hemophilia, treatment should also be provided.

### 2.2 PSYCHOSOCIAL ISSUES

Psychosocial factors have a significant impact on quality-of-life of patients with chronic diseases such as hemophilia. Interventions to support the psychosocial needs of patients and their carers, such as providing information and assistance, clarifying doubts, and teaching coping strategies to minimize the impact of the disabilities, may help to maximize the patient outcomes and improve the quality-of-life of their families.<sup>9,10</sup>

Programs and studies have shown positive results from psychological support to parents following the diagnosis of hemophilia. Parents who participated in a program of monthly psychological support and counselling showed significant progression, in pre- and post-study tests, in greater use of problem-focused coping strategies (recognition that a situation can change), and a reduced use of emotion-focused strategies (indicative of no change).<sup>11</sup>

#### RECOMMENDATIONS

People with hemophilia and their families should be provided with access to psychological and social support. Specifically, physicians should focus on patient and family education and addressing the emotional burden of the disease on the patient. Increased awareness of hemophilia is also needed amongst healthcare providers and the general population, for example through medical caravans and the media.



## 2. SPECIAL MANAGEMENT ISSUES

### 2.3 CIRCUMCISION

In Egypt, circumcision is an important cultural practice and must be considered in patients with hemophilia. Modern hemophilia treatment aims to integrate people with hemophilia in the society, acknowledging and respecting their culture and beliefs.

Such an approach is vital in order to succeed in hemophilia management in Egypt.<sup>12</sup>

Despite the high costs of the procedure and the risks of developing inhibitors, social demand for circumcision of boys with hemophilia is significant.<sup>12</sup>

Since the risks of developing inhibitors is higher during the neonatal period, postponing circumcision until after 6 months of age is advisable, and between 6–18 months is preferred.<sup>12,13</sup>

#### RECOMMENDATIONS

Circumcision of male patients with hemophilia and / or with a positive family history of hemophilia should not be considered a minor surgical procedure and must be performed under strict conditions. Clotting factor concentrate administration (even if minimal) is usually required before circumcision and should be continued during the following week of the procedure.

## 3. LABORATORY DIAGNOSIS

### 3.1 PRINCIPLES OF DIAGNOSIS

Early diagnosis is important for the effective management of hemophilia. A correct diagnosis is essential to ensure that a patient receives the appropriate treatment, particularly considering that different bleeding disorders may have very similar symptoms.<sup>1</sup>

Accurate diagnosis can only be made with the support of a comprehensive and accurate laboratory service. Screening tests should be used to identify the potential

cause of bleeding, for example, platelet count, bleeding time (in select situations) or other platelet function screening tests, prothrombin time (in select situations), and activated partial thromboplastin time (APTT).<sup>4</sup> Diagnosis should be confirmed by factor assays and other appropriate specific investigations. A central laboratory to confirm factor level is mandatory. Genotyping is essential, however, services are currently lacking in Egypt.

#### RECOMMENDATION

Specific WFH guidelines on laboratory practices for the diagnosis of hemophilia should be followed.

### 3.2 DETECTION OF INHIBITORS

“Inhibitors” in hemophilia refer to immunoglobulin G (IgG) antibodies that neutralize or limit the effectiveness of clotting factor concentrates.<sup>4</sup> Depending on the potency of inhibitors present, treatment may be rendered partly or completely ineffective, resulting in increased morbidity.<sup>14,15</sup> Inhibitor development is multifactorial, with genetic and non-genetic risk factors involved. Inhibitors occur in about 30% of previously untreated patients (PUPs) with severe hemophilia A,

usually within the first 50 to 100 exposure days (EDs) – but a baseline low risk remains throughout a patient’s life.<sup>16,17</sup> The impact of treatment-related factors on inhibitor development in PUPs with hemophilia A is still debated.<sup>18</sup>

Development of inhibitors in people with hemophilia is one of the most serious complications of hemophilia management today.<sup>1</sup>

#### RECOMMENDATION

Specific WFH guidelines on the detection of inhibitors should be followed.

## 4. HEMOSTATIC AGENTS AND DOSING REGIMENS

### 4.1 CLOTTING FACTOR CONCENTRATES

There are two categories of FVIII and FIX concentrate for the treatment of hemophilia A and B, respectively: plasma-derived (pd) and recombinant. Plasma-derived clotting factor is manufactured from large pools of human plasma, and undergoes various processes that aim to inactivate and eliminate known blood-borne viruses.

Recombinant clotting factor concentrates are manufactured from genetically engineered cell lines and undergo multiple purification steps, specific to product manufacturers.

### 4.2 PRODUCT SELECTION

Currently available therapeutic options for the management of hemophilia are highly effective. When selecting a coagulation factor, the main considerations are product safety and cost.

At a global scale, between 1970 and 1980, large numbers of people with hemophilia became infected with blood-borne viruses due to contaminated pdFVIII and pdFIX.<sup>19</sup> Careful selection of blood donors, screening of plasma, and advances in manufacturing of clotting factor concentrates have led to an almost complete elimination of the risk of known viral transmissions. However, despite the advances in prevention of transmission of known lipid-enveloped blood-borne viruses, blood can never be considered completely sterile as there are transitory or permanently circulating viruses that are not currently screened for, such as hepatitis E virus, Epstein-Barr virus, parvoviruses, cytomegaloviruses and Torque

Teno virus.<sup>20</sup> Additionally, non-lipid-enveloped pathogens may survive current viral inactivation processes, and emerging viral and non-viral pathogens, such as prions, should also be considered.<sup>21</sup>

The risk of prion-mediated disease through plasma-derived products is higher in the absence of a reliable screening test for variant Creutzfeldt-Jakob disease (vCJD), and when there are no established manufacturing steps to inactivate the vCJD prion.

It is also important to identify persistence of inhibitors against factor VIII, for example, as it may be a risk factor that increases physical disability in hemophilia A patients.<sup>22</sup> However, the development of neutralizing anti-FVIII inhibitors are not only dependent on the concentrate used.<sup>23</sup>

### RECOMMENDATIONS

Recombinant clotting factor concentrates are considered the gold-standard therapy and are the recommended treatment of choice for people with hemophilia in Egypt. If resources are limited, virally inactivated plasma-derived concentrates can be used. Due to the high prevalence of HCV in Egypt, cryoprecipitate, fresh frozen plasma and solvent and detergent filtration cryoprecipitate should only be used in emergency situations when no alternatives are available.

## 4. HEMOSTATIC AGENTS AND DOSING REGIMENS

### 4.3 PROPHYLAXIS VS ON-DEMAND THERAPY

Prophylaxis is the treatment of factor concentrate to prevent anticipated bleeding (Table 2).<sup>4</sup>

Prophylactic treatment has been shown to result in fewer hemarthroses, less arthropathy, fewer muscle bleeds, less frequent monitoring, fewer hospital admissions, reduced risk of cerebral bleeding, less joint surgeries, less time off work, lower disability and improved quality-of-life.<sup>24</sup>

It has been shown that annual factor usage may be similar in patients on prophylactic and on-demand treatment regimens.

Therefore, compared with a primarily on-demand treatment strategy, a primarily prophylactic treatment strategy can lead to better outcomes at equivalent treatment costs.<sup>23,25,26</sup>

### 4.4 CRYOPRECIPITATE

In Egypt, due to cost and resource constraints, both virally inactivated cryoprecipitate by solvent and detergent (S/D) and virally inactivated plasma by S/D are used for the management of patients with hemophilia.

Data have shown that S/D filtration cryoprecipitate FVIII “can be safely used for the control of acute and chronic bleeding episodes in hemophilia A patients”<sup>27</sup> but should only be used in “situations where clotting factor concentrates are not available”.<sup>2</sup>

Regarding dosage, the World Federation of Hemophilia highlights that “a bag of cryoprecipitate made from one unit of fresh frozen plasma (200–250ml) may contain 70–80 units of FVIII in a volume of 30–40 ml”.<sup>2</sup>

### 4.5 FRESH FROZEN PLASMA

Fresh frozen plasma (FFP) is a limited option to treat coagulation factor deficiencies as it is generally difficult to achieve FVIII levels higher than 30 IU/dl.<sup>2</sup>

Although FFP can be used for the treatment of hemophilia B in countries with limited resources to afford pdFIX concentrates, the WFH does not recommend its use due to its safety and quality concerns.<sup>2</sup>

**TABLE 2: Definitions of factor replacement therapy protocols<sup>1</sup>**

Protocol	Definition
<b>On-demand treatment (episodic)</b>	Treatment given at the time of clinically evident bleeding
<b>Primary prophylaxis</b>	Regular continuous* treatment initiated in the absence of documented osteochondral joint disease, determined by physical examination and / or imaging studies, and started before the second clinically evident large joint bleed and age 3 years**
<b>Secondary prophylaxis</b>	Regular continuous* treatment started after two or more bleeds into large joints** and before the onset of joint disease documented by physical examination and imaging studies
<b>Tertiary prophylaxis</b>	Regular continuous* treatment started after the onset of joint disease documented by physical examination and plain radiographs of the affected joints
<b>Intermittent prophylaxis (periodic)</b>	Treatment given to prevent bleeding for periods not exceeding 45 weeks in a year

\*Continuous is defined as the intent of treating for 52 weeks per year and receiving a minimum of an *a priori* defined frequency of infusions for at least 45 weeks (85%) of the year under consideration.

\*\*Large joints = ankles, knees, hips, elbows and shoulders.

## RECOMMENDATIONS

Prophylaxis is the preferred treatment approach for all patients to preserve musculoskeletal function, as it prevents bleeding and joint destruction. Prophylaxis is cost-effective in the long term because it eliminates the high cost associated with subsequent management of damaged joints and improves quality-of-life. Low-dose (recombinant, third generation 40 IU/kg)<sup>28</sup> 2–3 times per week prophylaxis is an effective option, but should be individualized according to age, venous access, bleeding phenotype, activity and availability of clotting factor concentrates. One option for the treatment of very young children is to start prophylaxis once per week and escalate depending on bleeding and venous access.

## 5. COMPLICATIONS OF HEMOPHILIA

### 5.1 SYNOVECTOMY

Approximately 80–90% of bleeding episodes in hemophilia occur in the musculoskeletal system, especially in the large synovial joints (elbows, knees, ankles, hips and shoulders).<sup>4</sup> Chronic hemophilic synovitis is characterized by persistent joint swelling and proliferative synovitis.<sup>29</sup>

A reduction of the hypertrophied synovium is the key to preventing recurrent intra-articular hemorrhages.<sup>29</sup> The vicious cycle of hemarthrosis–synovitis–hemarthrosis secondary to recurrent hemarthroses must be interrupted as soon as possible to avoid the development of hemophilic arthropathy.<sup>29</sup>

Synovectomy should be considered if chronic synovitis persists with frequent recurrent bleeding and it is not possible to be controlled by other means.<sup>1</sup> If a radioisotope is not available, chemical synovectomy with rifampicin is an appropriate alternative,<sup>1</sup> however, chemical synovectomy can be more painful than radioactive synovectomy.<sup>30</sup>

A combined prospective study comparing chemical (rifampicin) vs radioactive (<sup>90</sup>Y) synovectomy showed similar results with both methods.<sup>30</sup>

### RECOMMENDATIONS

Guided by ultrasound, non-surgical synovectomy is the procedure of choice. Chemical synovectomy should be used when appropriate and available. Rifampicin is highly effective, has few side effects, and can be accomplished in an out-patient setting, when preceded and followed by factor infusion, analgesic use and bed rest.

## 6. EGYPTIAN SOCIETY OF HEMOPHILIA AND FUTURE STRATEGIC OBJECTIVES

### 6.1 INTRODUCTION TO THE EGYPTIAN SOCIETY OF HEMOPHILIA (ESH)

The regular management of hemophilia in Egypt started through the establishment of a Diagnostic Unit of Inherited Bleeding Disorders (IBD) in the Central Health Laboratories of the Ministry of Health (MoH) and a Hemophilia Treatment Center in the National Institute of Diabetes, in 1968. Three years later, Dr Bothaina El Shenawy, together with a group of interested hematologists and hemophilia patients, founded the Egyptian Society of Hemophilia (ESH).

The ESH objectives are to:

1. Accurately identify IBD through the provided MoH laboratories and blood research departments;
2. Establish and maintain a national registry to accurately

determine the epidemiological data of IBD and improve decision-making on adequate 'patient' treatments;

3. Increase public awareness about IBD and advocate for patients with hemophilia rights in accessing appropriate healthcare and integration in the society through:
  - Free education
  - Exemption from military service
  - Employment rights similar to other impairment disorders

4. Scientific research in the field of IBD;

5. Cooperation with other national and international organizations that can help to improve care of patients with IBD.

### 6.2 NATIONAL CARE PROGRAM FOR IBD

As a result of the work by the ESH there are now about 21 hemophilia treatment centres across the country,

IBD are among government priorities and allocated budget is progressively increasing.

**Figure 1: Role of the Egyptian Society of Hemophilia**



## 6. EGYPTIAN SOCIETY OF HEMOPHILIA

### 6.3 ESH VISION AND STRATEGIC PLAN UNTIL 2018

#### 01 PUBLIC AWARENESS

There is interest in increasing knowledge about IBD among healthcare professionals and the community, particularly through governmental support

#### 02 PRODUCT AVAILABILITY

Increase the availability of FVIII from 0.36 IU/capita to 0.86 IU/capita

#### 03 NATIONAL GUIDELINES

Establish National Management Guidelines and ensure regular reviews and updates

#### 04 DIAGNOSIS

Introduction of a national screening program for inhibitors to FVIII and FIX

#### 05 NATIONAL REGISTRATION

Introduce regular evaluation of management outcome and establish a national registry

#### 06 COMMUNITY SUPPORT

Increase governmental support for rehabilitation of patients with IBD



## 7. BIBLIOGRAPHY

- World Health Organization. *Country Cooperation Strategy for WHO and Egypt 2010–2014*. Cairo, Egypt: World Health Organization Regional Office for the Eastern Mediterranean, 2010. Available from: [http://who.int/countryfocus/cooperation\\_strategy/ccs\\_egy\\_en.pdf?ua=1](http://who.int/countryfocus/cooperation_strategy/ccs_egy_en.pdf?ua=1) [Accessed October 2016].
- WFH Guidelines 2015. Available from: [www1.wfh.org/publication/files/pdf-1472.pdf](http://www1.wfh.org/publication/files/pdf-1472.pdf) [Accessed October 2016]
- Kabil N, ElAlfy MS, Metwalli N. Evaluation of the oral health situation of a group of Egyptian haemophilic children and their re-evaluation following an oral hygiene and diet education programme. *Haemophilia* 2007;13:287–92.
- Srivastava A, Brewer AK, Mauser-Bunschoten EP, Key NS, Kitchen S, et al. *Hemophilia* 2013;19:e1–e47.
- Lobet S, Hermans C, Lambert C. Optimal management of hemophilic arthropathy and hematomas. *J Blood Med* 2014;5:207–18.
- El-Zanaty F, Way A. *Egypt Demographic and Health Survey 2008*. Cairo, Egypt: Ministry of Health, El-Zanaty and Associates, and Macro International, 2009.
- Mohamoud YA, Mumtaz GR, Riome S, Miller D, Abu-Raddad LJ. *BMC Infectious Diseases* 2013;13:288.
- El Ekiaby M, Moftah F, Goubran H, van Drimmelen H, LaPerche S, et al. Viremia levels in hepatitis C infection among Egyptian blood donors and implications for transmission risk with different screening scenarios. *Transfusion* 2015;55:1186–94.
- Cassis FR, Querol F, Forsyth A, Iorio A, HERO International Advisory Board. Psychological aspects of hemophilia: a systematic review of methodologies and findings. *Hemophilia* 2012;18:e101–14.
- Tantawy A, von Mackenssen S, Mohammed AM, Labib JH, Moftah F, et al. Health-related quality of life in Egyptian children and adolescents with hemophilia A. *Pediatr Hematol Oncol* 2011;28:222–9.
- Bottos AM, Zanon E, Sartori MT, Girolami A. Psychological aspects and coping styles of parents with Haemophilic child undergoing a programme of counselling and psychological support. *Hemophilia* 2007;13:305–10.
- Zulfikar B, Karaman MI, Ovali F. Circumcision in Hemophilia: An Overview. 2003. Available from: [www1.wfh.org/publication/files/pdf-1170.pdf](http://www1.wfh.org/publication/files/pdf-1170.pdf) [Accessed October 2016].
- ElAlfy M, Elbarbary NS, Eldebeiky MS, El Danasoury AS. Risk of bleeding and inhibitor development after circumcision of previously untreated or minimally treated severe hemophilia A children. *Pediatr Hematol Oncol* 2012;29:485–93.
- Paisley S, Wight J, Currie E, Knight C. The management of inhibitors in hemophilia A: introduction and systematic review of current practice. *Hemophilia* 2003;9:405–17.
- Rivard GE, Rothschild C, Toll T, Achilles K. Immune tolerance induction in hemophilia A patients with inhibitors by treatment with recombinant factor VIII: a retrospective non-interventional study. *Hemophilia* 2013;19:449–55.
- EMA Guidance. EMA/CHMP/BPVWP/144533/2009. Available from: [http://ema.europa.eu/docs/en\\_GB/document\\_library/Scientific\\_guideline/2011/08/WC500109692.pdf](http://ema.europa.eu/docs/en_GB/document_library/Scientific_guideline/2011/08/WC500109692.pdf) [Accessed October 2016].
- Iorio A, Puccetti P, Makris M. Clotting factor concentrate switching and inhibitor development in hemophilia A. *Blood* 2012;120:720–27.
- Marcucci M, Mancuso ME, Santagostino E, Kenet G, ElAlfy M, et al. Type and intensity of FVIII exposure on inhibitor development in PUPs with hemophilia A. A patient-level meta-analysis. *Thromb Haemost* 2015;113:958–67.
- Di Minno G, Canaro M, Ironside JW, Navarro D, Perno CF, et al. Pathogen safety of long-term treatments for bleeding disorders: still relevant to current practice. *Haematologica* 2013;98:1495–98.
- Minno G, Canaro M, Ironside JW, Navarro D, Perno CF, et al. Pathogen safety of long-term treatments for bleeding disorders: (un)predictable risks and evolving threats. *Semin Thromb Haemost* 2013;39:779–93.
- Key NS, Negrier C. Coagulation factor concentrates: past, present, and future. *Lancet* 2007;370:439–48.
- Abdel-Messih IY, Habashy DM, Moftah SG, ElAlfy MS. Persistent factor VIII inhibitors and orthopaedic complications in children with severe haemophilia A. *Haemophilia* 2011;17:490–3.
- Peyvandi F, Mannucci PM, Garagiola I, El-Beshlawy A, ElAlfy M, et al. Randomized Trial of Factor VIII and Neutralizing Antibodies in Hemophilia A. *N Engl J Med* 2016;26:374:2054–64.
- Makris M. Prophylaxis in hemophilia should be lifelong. *Blood Transfus* 2012;10:165–8.
- Fischer K, van der Bom JG, Molho P, Negrier C, Mauser-Bunschoten EP, et al. Prophylactic versus on-demand treatment strategies for severe hemophilia: a comparison of costs and long-term outcome. *Hemophilia* 2002;8:745–2.
- Elekiaby A, El Saied H, Elekiaby M. Five years of tailored low dose prophylaxis in a small cohort of kids with severe hemophilia A using SD-F cryoprecipitate. WFH 2016 Congress, PO169
- El-Kiaby M, Goubran HA, Radosevich M, Abd-Allah A, El-Ekiaby A, et al. Pharmacokinetic study of minipooled solvent/detergent filtered cryoprecipitate factor VIII. *Haemophilia* 2011;17:1–5.
- BeneFIX Summary of Product Characteristics, 2016.
- Rodriguez-Merchan EC. Methods to treat chronic haemophilic synovitis. *Hemophilia* 2001;7:1–5.
- Rodriguez-Merchan EC, Caviglia HA, Magallon M, Perez-Bianco RI. Chemical synovectomy vs. radioactive synovectomy for the treatment of chronic haemophilic synovitis: a prospective short-term study. *Hemophilia* 1997;3:118–22.