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**INTRODUCTION**

The Clinical Practice Guidelines (CPG) on Management of Haemophilia was published in 2018. A Quick Reference (QR) and a Training Module (TM) are developed to increase the utilisation of the CPG. This TM has been developed by the members of Development Group (DG) of the CPG. The contents of the TM are extracted from the main CPG. It may be reproduced and used for educational purposes but must not be used for commercial purposes or product marketing.

**OBJECTIVES**

* To actively disseminate contents of the CPG and train healthcare providers on it; it may also be used for other educational purposes in the management of haemophilia in any healthcare settings in Malaysia
* To assist the ‘trainers’ in delivering all components related to the implementation of the CPG systematically and effectively

**TARGET USERS**

All healthcare providers involved in the management of haemophilia in primary, secondary and tertiary health care settings

|  |
| --- |
| This document contains a Training Module booklet on:   * Introduction, objectives, target users, authors and instructions for use * Proposed training programme/schedule * Test questionnaire * 10 lectures (in **PPT**) * 4 case discussions (in **PPT**) |

**AUTHORS**

Dr. Zulaiha Muda

Consultant Paediatric Haemato-oncologist

Institut Pediatrik/Women & Child Hospital (WCH)

Hospital Kuala Lumpur (HKL), Kuala Lumpur

**Members (alphabetical order)**

|  |  |
| --- | --- |
| Dr. Aisyah Muhammad Rivai  Consultant Paediatric Haemato-oncologist  Hospital Raja Permaisuri Bainun, Perak | Ms. Norhafizah Ayob  Physiotherapist  Institut Pediatrik/WCH, HKL, Kuala Lumpur |
| Dr. Azman Othman  Family Medicine Specialist  Klinik Kesihatan Tengkera, Melaka | Dr. Norjehan Yahaya  Specialist in Special Needs Dentistry  HKL, Kuala Lumpur |
| Dr. Cheah Yee Keat  Consultant Paediatrician  Hospital Tuanku Jaa’far, Negeri Sembilan | Dr Ong Gek Bee  Consultant Paediatric Haemato-oncologist  Hospital Umum Sarawak, Sarawak |
| Dr. Che Hadibiah Che Mohd Razali  Consultant Paediatric Hemato-oncologist  Hospital Sultan Ismail, Johor | Dr. Raja Zarina Raja Shahardin  Consultant in Paediatric Dentistry  Institut Pediatrik/WCH, HKL, Kuala Lumpur |
| Dato’ Dr. Goh Ai Sim  Senior Consultant Haematologist  Hospital Pulau Pinang, Pulau Pinang | Ms. Siti Mariam Mohtar  Senior Assistant Director  MaHTAS, MoH, Putrajaya |
| Dr. Kamalia Kamarulzaman  Nuclear Medicine Physician  Hospital Kuala Lumpur, Kuala Lumpur | Ms. Subasyini a/p Sivasupramaniam  Pharmacist  Institut Pediatrik/WCH, HKL, Kuala Lumpur |
| Dr Lim Soo Min  Consultant Haematologist  Hospital Sultanah Aminah, Johor | Dr. Wan Hayati Mohd Yaakob  Pathologist (Haematology)  Hospital Tuanku Ampuan Rahimah, Selangor | |
| Dr. Mohd Aminuddin Mohd Yusof  Head of CPG Unit &  Public Health Physician  MaHTAS, MoH, Putrajaya | Ms. Wong Shu Ping  Pharmacist  Hospital Ampang, Selangor |
| Mr. Mohd Helmi Hashim  Physiotherapist  Hospital Sultanah Bahiyah, Kedah | Dr. Yeoh Seoh Leng  Consultant Paediatric Haemato-oncologist  Hospital Pulau Pinang, Pulau Pinang |
| Dr. Nazzlin Dizana Din  Paediatric Haemato-oncologist  Hospital Sultanah Nur Zahirah, Terengganu | Dr.Yuslina Mat Yusoff  Pathologist (Haematology)  Institut Penyelidikan Perubatan, Kuala Lumpur |
| Miss Nor’Ashikin Johari  Consultant Paediatric Orthopaedic Surgeon  Institut Pediatrik/WCH, HKL, Kuala Lumpur |  |

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CPG Secretariat, Health Technology Assessment Section

Medical Development Division, Ministry of Health, Malaysia

4th Floor, Block E1 Parcel E, 62590 Putrajaya

E-mail: [**htamalaysia@moh.gov.my**](mailto:htamalaysia@moh.gov.my)

**INSTRUCTIONS FOR USE**

This Training Module consists of:

1. Lecture - ten sections
2. Case discussion - four sections
3. Training programme/schedule
4. Test questionnaire

(A booklet on this Training Module is enclosed together)

The training may be conducted in one day and consists of two parts. In part 1, didactic lectures are delivered to the whole group of training participants to inculcate the understanding on the management of haemophilia. In Part 2, participants are grouped into smaller groups to deliberate on cases of haemophilia with assigned facilitators. In both parts, there should be active participation from the training participants for effective learning.

The test questionnaire must be given to the training participants before the training session starts (pre-test) and after it ends (post-test). The pre-test is to assess the level of knowledge and understanding of training participants in the management of haemophilia. The post-test is to ascertain the increase in the training participants’ knowledge after attending the training session.

Should the trainers have any queries, kindly forward to [htamalaysia@moh.gov.my](mailto:htamalaysia@moh.gov.my)

**Training of Core Trainers on**

**CPG Management of Haemophilia**

|  |  |  |
| --- | --- | --- |
| **8 October 2019** | | |
| **Time** | **Lecture/case discussion** | **Lecturer/facilitator** |
| 0800 - 0830 | Registration  Pre-test | MaHTAS |
| 0830 - 0845 | Welcome & Introduction | Dr. Zulaiha Muda/Dr. Mohd Aminuddin Mohd Yusof |
| 0845 - 0900 | Clinical Presentation | Dr. Azman Othman |
| 0900 - 0930 | Laboratory Diagnosis | Dr. Wan Hayati Mohd Yaakob |
| 0930 - 1015 | Case Discussion 1 | Dr. Azman Othman/Dr. Wan Hayati Mohd Yaakob /Dr. Yuslina Mat Yusoff |
| 1015 - 1045 | **TEA BREAK** |  |
| 1045 - 1115 | Prophylaxis & Treatment for Acute Bleeding in Specific Sites | Dr. Ong Gek Bee/Dr. Che Hadibiah Hadibiah Che Mohd Razali |
| 1115 -1145 | Treatment of Musculoskeletal Complications | Miss Nor’Ashikin Johari |
| 1145 -1215 | Management of Inhibitors | Dato’ Dr. Goh Ai Sim |
| 1215 - 1300 | Case Discussion 2 | Dr. Nazzlin Dizana Din /Dr. Lim Soo Min |
| 1300 - 1400 | **LUNCH** |  |
| 1400 - 1430 | Non-pharmacological Treatment | En. Helmi Hashim |
| 1430 - 1500 | Home Therapy & Adherence/HMTAC | Pn. Wong Shu Ping/Pn. Subasyini a/p Sivasupramaniam |
| 1500 - 1600 | Case Discussion 3 | Dr Cheah Yee Keat/Pn. Norhafizah Ayob |
| 1600 - 1630 | **TEA BREAK** |  |
| **9 October 2019** | | |
| 0830 - 0900 | Special Situations | Dr. Aisyah Muhammad Rivai |
| 0900 - 0930 | Dental Care | Dr. Raja Zarina Raja Shahardin |
| 0930 - 1015 | Case Discussion 4 | Dr. Norjehan Yahaya /Dr Aisyah Muhammad Rivai |
| 1015 – 1045 | **TEA BREAK** |  |
| 1045 – 1115 | Monitoring | Dr. Yeoh Seoh Leng |
| 1115 – 1200 | Post-test & Closing | Dr. Cheah Yee Keat/Dr. Zulaiha Muda |
| 1200 | **LUNCH** |  |

**TEST QUESTIONNAIRE**

**Answer all questions by circling the right answers.**

| **No.** | **Questions** | **Answer** | |
| --- | --- | --- | --- |
| **True** | **False** |
| **1.** | **The following statements are true.** | | |
| 1. Spontaneous intracranial bleed in newborn can be due to haemophilia. | T | F |
| 1. A positive family history is present in one-third of patients while another two-third may have spontaneous mutation. | T | F |
| 1. Bleeding in the throat is considered life-threatening in haemophilia. | T | F |
| 1. Severe haemophilia A is defined as FVIII level <5%. | T | F |
| 1. Spontaneous bleeding is common in mild haemophilia. | T | F |
|  | | | |
| **2.** | **With regards to the following laboratory investigations in haemophilia, which statements is/are true?** | | |
| 1. Factor VIII deficiency will cause isolated prolonged activated partial thromboplastin time (APTT). | T | F |
| 1. Mixing test is required when APTT showed normal result. | T | F |
| 1. Platelet count is reduced in haemophilia A. | T | F |
| 1. Bethesda test of 0.5 BU is considered negative. | T | F |
| 1. Intron 22 mutation is common mutation in haemophilia B. | T | F |
|  | | | |
| **3.** | **With regards to clotting factor concentrates (CFC), the following is/are true.** | | |
| 1. CFC can be plasma derived or recombinant. | T | F |
| 1. When there is no improvement within 12 hours or worsening of symptoms requiring >2 infusions for complete resolution, it is considered a poor response. | T | F |
| 1. When starting a patient with any type of CFC, virology studies should be done regularly. | T | F |
| 1. On demand therapy is able to reverse the deleterious effects of bleeding on synovial tissues. | T | F |
| 1. Prophylactic factor replacement therapy is defined as regular infusion of CFC in an attempt to raise clotting factor levels and to keep them at 1% or higher at all times. | T | F |
|  | | | |
| **4.** | **A 5-year-old boy of haemophilia A presents with haematuria. Which of the following statements are true?** | | |
| 1. Vigorous hydration should be started at 3 L/m2 for a minimum of 6 hours. | T | F |
| 1. Initiate factor replacement within 2 hours. | T | F |
| 1. Start tranexamic acid/antifibrinolytic therapy to control the bleeding | T | F |
| 1. Factor level needs to be raised up to 50% if there is pain or persistent gross haematuria after 48 hours. | T | F |
| 1. Refer urologist if urinary tract obstructions is suspected. | T | F |
|  | | | |
| **5.** | **Haemathrosis:** | | |
| 1. is the most common bleeding manifestation in haemophilia | T | F |
| 1. for treatment, it is required to raise the factor level up to 40 - 60% | T | F |
| 1. with hip joint involvement is common | T | F |
| 1. is referred for physiotherapy once recovered | T | F |
| 1. needs urgent referral to orthopedic for knee aspiration | T | F |
|  | | | |
| **6.** | **The following is/are true regarding inhibitor in haemophilia.** | | |
| 1. Screening for inhibitor should be done within four weeks of intensive treatment. | T | F |
| 1. rFVIIa at a dose of 90 - 120 ug/kg every 2 - 3 hours is used to treat acute bleeding. | T | F |
| 1. Immune tolerance induction should not be considered for adult haemophilia patient who has inhibitor for many years. | T | F |
| 1. On demand therapy has lower incidence of inhibitor development compared with prophylaxis. | T | F |
| 1. Inadequate response to factor replacement therapy should prompt screening for inhibitor. | T | F |
|  | | | |
| **7.** | **Non-pharmacological treatment in persons with haemophilia (PWH):** | | |
| 1. PWH with acute knee haemarthrosis are encouraged to weight bear immediately to prevent contracture. | T | F |
| 1. “PRICE” is an important measure in acute pain management. | T | F |
| 1. Rehabilitation, e.g. hydrotherapy, mechanical exercises and strengthening exercises, improves joint health status. | T | F |
| 1. PWH are encouraged to participate in high contact sports to improve their self-esteem. | T | F |
| 1. PWH with high BMI are associated with increased arthropathic pain and risk of developing target joints. | T | F |
|  | | | |
| **8.** | **With regards to circumcision in PWH:** | | |
| 1. Circumcision is considered life-threatening and hence it is not obligatory by Muzakarah Jawatankuasa Fatwa Majlis Kebangsaan Bagi Hal Ehwal Ugama Islam Malaysia Kali Ke-77. | T | F |
| 1. Circumcision is absolutely contraindicated in PWH. | T | F |
| 1. If circumcision is indicated, it should be done in a haemophilia treatment centre. | T | F |
| 1. PWH with inhibitors may develop more serious complications after circumcision. | T | F |
| 1. Adjunctive antifibrinolytic therapy should be used with factor if circumcision is indicated. | T | F |
|  | | | |
| **9.** | **In person with haemophilia,** | | |
| 1. comprehensive oral health care should be initiated early within six months after the first tooth erupts | T | F |
| 1. NSAIDs is the best analgesic after dental extraction | T | F |
| 1. dental examination conducted when indicated | T | F |
| 1. brushing teeth should be avoided to prevent bleeding | T | F |
| 1. dietary counselling should be advocated to prevent dental diseases | T | F |
|  | | | |
| **10.** | **Monitoring measures of PWH are:** | | |
| 1. Annual bleeding rate | T | F |
| 1. Yearly magnetic resonance imaging | T | F |
| 1. Weekly inhibitor screening | T | F |
| 1. Annual Haemophilia Joint Health Score | T | F |
| 1. Proper documentation of factor administration: dose and response | T | F |

**ANSWERS FOR TEST QUESTIONNAIRE**

|  |  |  |  |  |  |  |  |  |
| --- | --- | --- | --- | --- | --- | --- | --- | --- |
| **Question** | | **Answers** | **Question** | | **Answers** | **Question** | | **Answers** |
| **1.** | a. | **T** | **5.** | a. | **T** | **8.** | a. | **T** |
| b. | **F** | b. | **T** | b. | **F** |
| c. | **T** | c. | **F** | c. | **T** |
| d. | **F** | d. | **F** | d. | **T** |
| e. | **F** | e. | **F** | e. | **T** |
| **2.** | a. | **T** | **6.** | a. | **T** | **9.** | a. | **T** |
| b. | **F** | b. | **T** | b. | **F** |
| c. | **F** | c. | **F** | c. | **F** |
| d. | **T** | d. | **F** | d. | **F** |
| e. | **F** | e. | **T** | e. | **T** |
| **3.** | a. | **T** | **7.** | a. | **F** | **10.** | a. | **T** |
| b. | **T** | b. | **T** | b. | **F** |
| c. | **F** | c. | **T** | c. | **F** |
| d. | **F** | d. | **F** | d. | **T** |
| e. | **T** | e. | **T** | e. | **T** |
| **4.** | a. | **F** |  |  |  |  |  |  |
| b. | **F** |  |  |  |  |  |  |
| c. | **F** |  |  |  |  |  |  |
| d. | **T** |  |  |  |  |  |  |
| e. | **T** |  |  |  |  |  |  |