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Needs of patients with hemophilia: Suggested clinical pathway

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Hemophilia is a worldwide hereditary bleeding disorder characterized by decreased function or absence of factor VIII or factor IX. Patients with hemophilia have physical, psychological and social needs.Therefore, the aims of the current study weretoexplore needs of patients with hemophilia and to suggest clinical pathway for these patients. To fulfill the aim of this studyanexploratory descriptive research design was utilized. A purposive sample of 30 patients with hemophilia was selected over a period of 3 months from different hematology units at governmental hospitals.Two tools were utilized (I) personal and medical data, (II) needs assessment of patients with hemophilia. The study findings revealed that,(43.3%) of the studied patients had hepatitis C virus and (26.6%) of them had deformity in the shape and functions of knees joints. (40%) of the studied patients received information about their disease from other patients. (90%) of the studied patients mentioned that;themedical team did not receive patients when they were admitted to the hospital and all studied patients stated that; there were no one of medical team did hospital orientation for them, no one of them engage the patients in their treatment plan and all studied patients did not perform any kind of exercises or perform pleasure activities and (90%) of them did not find the medical team when needed. (96.7%) of the studied patients were not satisfied with the medical services provided to them.Based on the results of the current study it could be concluded that; patients with hemophilia have physical, psychological and social needs. A comprehensive multidisciplinary assessment of the physical, emotional, and social needs of adult hemophilic patients is essential. Clinical pathway may provide comprehensive health care for patients with hemophilia based on patients 'needs to ensure that all important issues are involved also clinical pathway has been recommended to improve clinical outcomes, decrease mortality rates, and improve cost-effectiveness in health care settings.
KEY WORDS:*needs, patients with hemophilia, clinical pathway*

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I. Background

Hemophilia is the most common inherited bleeding disorder characterized by decreased function or absence of factor VIII (classic hemophilia or hemophilia A) or factor IX (Christmas disease or hemophilia B).Hemophilia is a male disorder, yet females with hemophilia are rarely seen in communities but they are carriers. Hemophilia A and B are inherited as X-linked genetic recessive disorder but 30% of patients have no family history of the disease and they usually have spontaneous new mutation(Christmas disease or hemophilia B)(Mulliez, Vantilborgh, Devreese, & June 2014 and Kobl, 2018).

Hemophilia A is four times more common than hemophilia B,this account for about 80 percent of the total number of patients with hemophilia, and about 70 percent of them have the severe form. Hemophilia affects people from all races. Hemophilia as a lifelong disorder with three phenotypes (severe, moderate and mild) that correlate with factor VIII (FVIII) levels in plasma. From 5 to 40 percent of normal clotting factor is considered mild, 1 to 5 percent is moderate, and less than 1 percent is severe.Absence of clotting factors increase bleeding time after minor injury or cause spontaneous bleeding in patients with severe disease especially in patients' joints. Contrary to popular belief, people with hemophilia do not bleed faster they bleed for a longer period of time(Souza, et al 2016; Crosta, 2017 & Centers for Disease Control and Prevention, 2018).

Complications of hemophilia may result either from the disease process or from lack of patients' information about the disease and how to deal with it. These complications include physical such as deep internal bleeding, 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). Chronic hemophilic synovitis is characterized by persistent joint swelling and proliferative synovitis, it may cause joint arthropathy, and loss of mobility. Other bleeding episodes can occur at any part of the body such as hematuria, epistaxis, and bleeding from the mouth or tongue. Bleeding into the head, neck, abdomen or gastro-intestinal tract is considered life-threatening and must be treated as an emergency(El-Beshlawy, et al 2016 &Hemophilia symptoms and causes, 2019).

Otherphysicalcomplications that may occur for patients with hemophilia includejoints damage, infection from blood or blood product transfusion such as hepatitis B, C and AIDS, adverse reaction to

clotting factor treatment or blood product infusion, antibody formation to factor products known as inhibitors.Additionally, psychosocialcomplications that may occur including fear, anxiety, stress, depression, lack of interest, social isolation, and inability to perform role, and feeling of dependency (Australian Haemophilia Nurses Group, 2013).

Diagnosis of hemophilia involves consideration of the patient's and family history, physical examination and testing the blood for its ability to clot and its levels of clotting factors (factor assays) (Butler, 2012 & Centers for Disease Control and Prevention, 2018). Hemophilia is treated with replacement therapy (Clotting factors) that can be derived from human blood (Clotting factor concentrates) or synthetically produced in a laboratory (recombinant clotting factors) which is considered the treatment of choice to reduce the risk of transmitting infections that are carried in human blood. Some patients will need regular replacement therapy (prophylactic therapy) in order to prevent bleeding. Others receive demand therapy that is given only after bleeding begins and remains uncontrollable (Shiozawa , 2015&Crosta, 2017).

Patients with hemophilia have physical, psychological and social needs, these needs include; information about their disease and how to deal with it, physiotherapy, psychological and social support and counseling, nutritional advice, medication regimen and follow-up, home therapy, and occupational advisor. Hematology nurses play a crucial role in providing comprehensive care to these patients to promote optimal physical, psychosocial and emotional health, and to minimize complications from bleeding and treatment as well as improve quality of life while decreasing morbidity and mortality rate (Breakey, Blanchette&Bolton-Magg, 2010; Morfini, et al 2015&Lazure, et al 2019).

Nursing responsibilities include assessing patients and their families' needs, collaborate with all health team members to formulate comprehensive care plan for these patients, providing direct clinical care, patient and family support, providing clear patient education about their condition, and administering treatment. Also, hematology nurse is responsible for providing assistance during lifestyle adjustments, and monitoring for early signs of complications, assessing the severity of bleeding episodes, helping families to select treatment options, selecting candidates for prophylaxis or home therapy, and carefully observing clinical progress for the patients. (Srivastava, et al 2012& El-Beshlawy et al, 2016).

The World Federation of Hemophilia (WFH), (2017) recommends that; the wide range and changing needs of patients and their families are best met through a multidisciplinary comprehensive care team that includes hematologist, hematological nurse, surgeon, physical therapist, psychologist, nutritionist, and social worker, with access to additional specialists as needed in accordance with accepted protocols that are practical and national treatment guidelines or clinical pathway.Suggested clinical pathway provides comprehensive health care for patients with hemophilia and their families based on patients 'needs to ensure that all important issues are involved. Moreover, clinical pathway has been recommended to improve clinical outcomes, decrease mortality rates, and improve cost-effectiveness in health care settings. (Mansour, 2015 and Breakey, Blanchette&Bolton, 2010). Therefore, the aim of the current study is to explore needs of patients with hemophilia and suggest clinical pathway for their care.

Significance of the study:

World widely, there are more than 400,000 people with hemophilia;about1 in 5,000 males has hemophilia(Centers for Disease Control and Prevention, 2018). According to the World Federation of Hemophilia (WFH), the estimated number of hemophilicpatients in Egypt is around 7500,000 (WFH, 2015), Another search concluded that; in Egypt around 8,300 patients with hemophilia (The First Center of Excellence for Hemophilia Treatment Launches in AtfalMisr Health Insurance Hospital, 2018).Hemophilia is a serious lifelong disease, and patients must depend on infusions of clotting factors several times per week to prevent bleeding throughout their lives, although effective treatments do exist, but they are expensive (Zhao, Zhao, Wang, Guo, & Wu, 2017 & National Hemophilia Foundation, 2019).

In fact; patients with hemophilia can be hospitalized by severe spontaneous life threatening episodes of bleeding. The bleeding can be fatal if it cannot be stopped or if it takes place in the brain or another vital organ. Moreover, hemophilic patients are prone to manycomplications. hemophilia not only affects patients by its clinical consequences, but also causes a great economic problem to health care systems due to costs of frequent hospitalizations, follow up visits and drugs in addition to costs due to decreased productivity of the affected individuals at work and schools therefore,the need for comprehensive care is very necessary for these patients. The comprehensive management plan should be individualized and adapted in accordance with patients' changing needs.

Review of literature shows few nursing studies about hemophilia also about developing of clinical pathway for these patients. although two recent studies were conducted to examine the impact of clinical pathway on patients' outcomes revealed that clinical pathway proved to be an effective approach to decrease physical complications, hospital cost, and length of hospital stay and improve patients' outcomes (Liang, Jun, Jun &Feng, 2018 and Mauricio, Moreno & Bonilla, 2019). Moreover, during the clinical experiences the

researchers observed thatmostof health team member not provide information to patients with hemophilia. Yet there is scarcity of researches applied to study the needs of patients with hemophilia also there is little researches that developed clinical pathway for these patients in Egypt.

Therefore, this study could be beneficial in providing health care providers especially nurses with data base regarding the needs of patients with hemophiliathat utilized by health care professional as well as suggest clinical pathway for these patients to maintain cost effectively and reduce unnecessary delays in care, decrease mortality rate for patients with hemophilia.Also, this study might have a positive reflection on patients' general health status during and after hospitalization, through improving patients' quality of care by reducing the physical and psychological complications. Furthermore, this study may contribute some evidential data to help health team members specially hemophilia nurse to integrate it in the care of patients with hemophilia. Data derived from this study may provide a nucleus for further qualitative and quantitative studies in this field.

Aim:

The aims of this study were to**:**

- Explore the needs/problems of patients with hemophilia.
- Suggest clinical pathway for patients with hemophilia.

The research questions:

To fulfill the aims of the current study the following research questions were formulated:

- What are the needs of patients with hemophilia?
- What is the suggested clinical pathway for patients with hemophilia?

Research design:

An exploratory, descriptive research design was used in the current study.

Setting:

This study was conducted in different hematologyunitsatgovernmental hospitals. Theseunits were established to provide treatment to all types of blood diseases for adults.

Sample:

A purposive sample consisting of 30 patients with hemophilia were included. This number was calculated using a power analysis and recruited to this study according to the following inclusion criteria.

a) Conscious adult male patients.
b) Had an age range between 18 to 55 years.
c) Able to communicate through talking.
d) Free from mental problems or any cognitive impairments that would prevent them from sharing their self-experiences
e) Agreed to participate in the study.
f) Able to sign a consent form.

Tools of Data Collection

The researchers used two tools to assess need of patients with hemophilia which include; personal and medical data (tool 1),and needs of patients with hemophilia (tool 2).

1)Personal data andMedical

Part one (personal data)

It is designed to collect data specific to each patient, which included; patient's code, age, gender, place of residence, level of education, marital status, andtype of work.

Part two (medical data)

This part included patient's medical data such as type of hemophilia, the first time the symptoms of disease were discovered, presence of comorbid diseases, common sites of bleeding, physical and psychological complications that developed from hemophilia.

2) Needs assessment of patients with hemophilia

This tool included questions to assess patients' needs and their suggestions to improve quality of their care which include main source of patients' information about disease, availability of health team members when needed, availability of patients' orientation for hospitalization, application of exercises during

hospitalization, performing leisure activities during hospitalization, patients' satisfaction about the care provided to them and their suggestion to improve care provided to them.

Tools Validity and Reliability

Content validity of the study tools were tested by panel of fiveexperts. Three expertswere faculty members in Medical Surgical Nursing field from Faculty of Nursing Cairo University and two experts were from hematology field. The experts were asked to examine the tools for content coverage, clarity, wording, length, format, and overall appearance. Reliability of the study tools were tested using test-retest (0.8).

Pilot Study

A pilot study was conducted on 10 patients with the same inclusion criteria to ensure the feasibility of the study and the study tools for data collection, as well as to examine issues related to the research design, time required to fill out the sheet, data collection procedures, any possible problems in the methodological approach and data analysis approaches. Based on feedback taken from the pilot study there were no modifications done in the study tools. The pilot sample was included in the study sample.

Ethical consideration:

An official permission was obtained from hospitals administrators to conduct the study. The purpose and nature of the study as well as the importance were explained to the patients who met the inclusion criteria. Signed consent was obtained from the patients who choose to participate in the study. Also, anonymity and confidentiality were assured through coding the data. Patients were assured that participation in this study was voluntary and they have the right to withdraw from the study at any time without penalty.

Procedure:

The study was conducted on four phases; assessment, planning, implementation, and evaluation phase. Assessment phase involves collecting data through reviewing the literature dating back to at least 5 years using scientific data base**.** Planning phase; based on the outcome of the assessment phase, final format of the study tools were developed. Final decision about time, frequency, of patients' interview was develop.Implementationphase;firstly, the researchers communicated with the head nurses of different haematology units to identify patients with haemophilia. Then; the researchers conducted an initial meeting with each patient individually for 10 minutes, in this first patient contact, the researchers introduced themselves to the patient, explained the nature of the study and took the initial agreement from the patient to participate in the study. Then; each patient who were met the inclusion criteria for the study was approached individually for 30 – 45 minutes by the researchersto collect personal, and medical information (tool 1) as well as to collect data about needs of patients' with haemophilia (tool 2).Data was collected over a period of 3 months.Evaluation phase; after finishingthe collected data were analysed statistically to assess needs of patients with haemophilia.Based on results of previous data and reviewing the literature dating back to at least 5 years using scientific data base, suggested clinical pathway for patients with hemophilia was developed.

Data analysis:

Collected data was tabulated, computed, and analyzed using SPSS, version 20. Descriptive statistics including frequency distribution, percentage, means and standard deviationswere used.

Presentation and analysis of data

Statistical findings of the current research are presented in two main sections as follows:

Section I: Descriptive statistical findings of the studied patients related to personal and medical information, (table 1 - 3).

Section II: Descriptive statistical findings related to needs of studied patients and their suggestions to improve medical care provided (table 4).

Section I: Descriptive statistics findings related to personal and medical information of the studied patients (Table 1 - 3).

Table (1) illustrated that; almost of two thirds of the studied patients are in age range between 28 to less than 38 years,the mean age was (33.66 ± 0.583). All of the studied patients are male, 63.3% are married and 10 % divorced, 46% of them are self-employed and 26.7% have office work. Eighty percent of them reside in urban areas.

Table (1): Frequency and percentage distribution of patients' personal information (n. = 30).

Item	No.	%

			SD ± X
Age			
- 18 – < 28 years	4	13.3	
-28 – < 38years	20	66.7	33.66 ± 0.583
-38 – < 48 years	6	20	
Gender			
-Male	30	100	
-Female	0	0	
Marital status			
-Married	19	63.3	
- Single	8	26.7	
-Divorced	3	10	
Education			
- University degree	8	26.7	
-Secondary school diploma	9	23.3	
-Can read and write	7	16.7	
-Can't read or write	6	20	
Type of work			
- Office work	8	26.7	
-Self-employed	14	46.7	
-Student	3	10	
- Unemployed	5	16.6	
Place of residence			
-Urban	24	80%	
-Rural	6	20%	

Table (2) described that; 60% of the studied patients showed positive history of consanguinitytype of hemophilia, and 90%ofthem had type (A) hemophilia. All patients reported that; the first time the symptoms of the disease was discovered by uncontrolled bleeding at circumcession.As for comorbideties, 26.6% and 16.7% of the studied patientss reported having diabetes mellitus and renal stones respectively. Common sites of bleeding, 36.8% and 30% of the studied patient were had hemarthroses and excessive post traumatic bleeding respectively.

Table (2): Frequency and percentage distribution of studied patients' medical information (n. = 30).

Item	No.	%
Consanguinity		
- Positive	18	60
- Negative	12	40
Type of hemophilia		
A	27	90
B	3	10
The first time the symptoms of the disease were discovered		
-Uncontrolled bleeding during circumcision	30	100
Comorbidities		
- Hypertension	4	13.3
-Diabetes mellitus	8	26.6
-Renal stones	5	16.7
-Peptic ulcer	2	6.7
-Intestinal obstructions	1	3.3
- No	10	33.4
Common sites of bleeding		
-Hemarthroses	11	36.8
-Epistaxis	5	16.7
-Excessive post traumatic bleeding	9	30
-Hematuria	2	6.6
-Oral bleeding	2	6.6
-Cephalic bleeding	1	3.3

Table (3) shows that; in relation to question related to the physical complications caused by the disease, 43.3% of the studied patients reported havinghepatitis C virus and 26.6% of them had deformity in the shape and functions of knees joints. According to question asking about psychological, social and financial problems caused by the disease, 30% of the studied patients reported increase work absenteeism and insufficient income.

Table (3): Frequency and percentage distribution of patients' medical information (n. = 30).

Item	No.	%

Physical complications caused by the disease		
-Deformity in the shapes and functions of knees joints	8	26.6
-Deformity in the shapes and functions of elbow joints	4	13.4
-Deformity in the shapes and functions of ankle joints	3	10
-Atrophy in calf muscles	2	6.7
-Hepatitis C virus	13	43.3
Psychological, social and financial problems caused by the disease		
-Feeling of loneliness and insecurity	5	16.7
-Inability to do social activities and play the role	7	23.3
-Unavailability of clotting factors medications	7	23.3
-Increase work absenteeism and insufficient income.	9	30
-Inability to fulfill the educational study requirements	2	6.7

Section II: Descriptive statistical findings related to needs of studied patients and their suggestions to improve medical care provided (4).

Table (4) illustrated that; 40% of the studied patients receive information about hemophilia from other patients as a main source while only6.7% of them received information from heath team members. According to question about the patients perform any kind of hobbies/ leisure activities while they were in the hospital 100% of the studied patients their answered were no and 100% of them answered that, because there were not any one encourage them. Ninety percent of the studied patients answered no about if they were received by any one of the medical team upon admission to the unit. All patients answered no for questions relate to getting unit orientation by medical team, being engaged in their treatment plan, helped to perform any kind of exercises and 100% of them answered that, because there were no one help them to do exercise.

More than ninety percent of the studied patients were not satisfied with the medical services provided to them. Regarding to question asking about the studied patients' suggestions to improve medical services 40% of them suggested that, raising health team members and community members' awareness about the disease and 33.3% of them suggested to establish specialized hospitals to provide comprehensive care for patients with hemophilia and only 6.7% of them suggested to use new modalities to treat hemophilia such as gene therapy.

Table (4-1): Frequency and percentage distribution related to needs of studied patients and their suggestions to improve medical care provided

Item	**No.**	**%**
What is the main source of your information about hemophilia?		
-Health team members	2	6.7
- Internet	8	26.6
-Reading	2	6.7
-Other patients	12	40
-Medical Charities	6	20
Did the medical team receive you when you were admitted to the hospital?		
- Yes	3	10
-No	27	90
Did any of the medical team orient you with the unit?		
-Yes	0	0
-No	30	100
Did the medical team engage you in your treatment plan?		
-Yes		
-No	0	0
Did you perform any kind of exercises while you are in the hospital?	30	100
-Yes	0	0
-No	30	100
Why?No one helped me to do so	30	100
Did you perform any kind of hobbies/leisure activities while you are in the hospital?		
-Yes	0	0
-No	30	100
Why?Because there is no one to encouraged me to do so.		
Do you find the medical team when needed?	30	100
-Yes		
-No	3	10
	27	90

Table (4 - 2): Frequency and percentage distribution related to needs of studied patients and their suggestions to improve medical care provided

Item	No.	%
Are you satisfied with the medical services provided to you?		
Yes	1	3.3
No	29	96.7
What are your suggestions for improving medical services?		
-Raise health team members and community members awareness about the disease.	12	40
-Availability of clotting factors medication.		20
-Establish specialized hospitals to provide comprehensive care for patients with hemophilia.	6	33.3
-Use new modalities to treat hemophilia such as gene therapy.	10	
	2	6.7

II. Discussion

Section I:

Hemophilia is a life-long disorder which creates profound physical, emotional, economic and social problems for those patients. These problems extend beyond the patients to their families, their acquaintances, and society in general. In the present study, all the studied patients were males and eighty percent of them, are below thirty eight years of age. These results were in concordant with the results published by (Tawfiket al, 2017) findings of hemophilic patients in Assiut Egypt, also, the study done by (Valizadeh, et al 2014 &Tonbaryet al, 2010) in their study concluded that; all patients in two previous studies were male and the majority of them diagnosed with hemophilia in young age, the last study added that; early, being a young adult with hemophilia has its challenges. They often struggle with the stress of accepting responsibility for their hemophilia care and poor decision-making may lead to poor compliance that affects hemophilic patients' lifestyle.

In the current study, the majority of diagnosed hemophilia patients,ninety percent were hemophilia type A and only ten percent were hemophilia type B. These results were in concordant with the results of study conducted by (Speybroeck, & Hermans, 2015) about "Participation of people with hemophilia in clinical trials of new treatments: an investigation of patients' motivations and existing barriers" found that the majority of hemophilia cases eighty percent were hemophilia type A.

Positive consanguinity in the studied patients was present in sixty percent while negative consanguinity represented in forty percent. These results were in agreement with the results of (Al-Zubaidy, 2014) who carried descriptive study on patients with hemophilia in Al-Ramadi city, as positive consanguinity were reported in more than seventy percent and negative consanguinity in more than twenty five percent of hemophilic patients. Also these results were in agreement with the results of the study done by (Borhany et al, 2010) "Bleeding disorders in the tribe: result of consanguineous in breeding." Positive consanguinity marriage in Pakistan wasmore than sixty percent and negative consanguinity were more than thirty five percent of hemophilic patients. In the present study. All the studied patients reported that; the first time the symptoms of disease were discovereduncontrolled bleeding after circumcision. This result was in agreement with study conducted in Mansoura, Egypt by (Tonbaryet al, 2010) who reported that; the common presenting symptom was bleeding following male circumcision.

Regarding the presence of other diseases/health problems, two third of the studied patients were had co-morbid diseases such as diabetes mellitus, renal stones and hypertension (more than twenty five percent,more than fifteen percent&more than ten percent) respectively. This finding is consistent with a study carried by **(**Treil, 2014) aimed to assess physical and psychosocial challenges in adult hemophilia patients with inhibitors. who reported that; patients with hemophilia may experience other comorbid conditions such as diabetes, hypertension, renal disease, and cancer and who added that; these challenges may complicate the management of adult patients further are associated with increased bleeding symptoms and clotting factor consumption.

In the present study, more than forty percent of the hemophilic patients had hepatitis C in comparison with the results of the previous studies done by (Tawfik, el al, 2017) who found positive HCV in more than five percent of studied hemophilic patients. Also (Tonbary et al, 2010) in their study reported that; hepatitis C infection and arthropathy represented the complications in patients with hemophilia and they added that; unfournitly, those patients are potential risks underlying the use of blood products. Although, recent advances in blood product processing and its applications have virtually eliminated the risk of transmission for most pathogens, including HIV, and hepatitis B and C. However, the safety of blood products can never be completely guaranteed (HoseiniValizadeh, Zamanzadeh, Fallahi, Behtash& Mina. 2014).

Based on literature review, 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. Additionally, 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.

Regarding common sites of bleeding, the finding of this study revealed that more than thirty five percent and thirty percent of the hemophiliac patients were had bleeding in knees joints and excessive post traumatic bleeding respectively. The results of the present study are congruence with study carried by (Knobe & Berntorp, 2018) entitled as hemophilia and joint disease reported that; bleeding most commonly occurs in the knees, elbows, and ankles, and is often evident from early childhood. Recurrent joint bleeding causes synovial proliferation and inflammation (haemophilicsynovitis) that contribute to end-stage degeneration (haemophilicarthropathy); with pain and limitation of motion severely affecting patients' quality of life.

The most common physical complications detected in the current study were deformity in the shapes and functions of knee and elbow joints which represented in more than twenty five percent andmore than ten percent respectively of all studied hemophilic patients compared with more than five percent of them complaining from atrophy in calf muscles. These results were in concordant with that of (Tonbary et al, 2010) who reported that; hemarthrosis was the most common complication among their hemophilic patients, also (Tawfik, el al, 2017) in their study reported that; the most common complications were hemarthrosis in more than twenty five patients, muscle hematoma in more than fifteen percent. Also, they added that increased complications in patients received on demand treatment compared to those received prophylaxis treatment.

Concerning Psychological, social and financial problems caused by the disease, the finding of this study revealed that; the high incidence of work absenteeism and insufficient income, unavailability of clotting factors medications, inability to do social activities and play the role, feeling of loneliness and insecurity unable to achieve educational requirements. These findings were in agreement with study done by (Treil, 2014) who reported that; numerous challenges confront adult hemophilia patients with hemophilia such as, deterioration of joints, physical disability, emotional turmoil, and social issues. Moreover, treatment regimens often used in the treatment of patients with hemophilia impose significant scheduling, economic, and emotional demands on patients and their families or primary caregivers. Patients have higher incidences of mobility-related problems, hospitalizations, school and work absenteeism, and difficulty maintaining a job, along with higher treatment costs.

In the same line, (Valizadeh, el al, 2015) carried study entitled as "Practice of Iranian Adolescents with Hemophilia in Prevention of Complications of Hemophilia" and reported that; although the complications of hemophilia result in numerous psychosocial outcomes, to the best of our knowledge, no previous study has evaluated these complications and their prevention methods**.**

Section II:

The current study showed that,forty percent of the studied patients the main source of patients' information about hemophilia was from other patients while less than percent of them received information from heath team members. In comparison with (Cutilli et al, 2010)who conducted study entitled as "Seeking health information: what sources do your patients use" and reported that;the most common and trusted source of information for patients is healthcare professionals, and they added that; patients use other sources of health information (e.g., TV, Internet, and family/friends/coworkers) to supplement information provided by healthcare professionals. Nurses should utilize knowledge about hemophilia to assist patients in obtaining health information to optimize health outcomes.

In relation to needs of the studied patients from health care providers, the majority of patients ninety percent stated that; medical team didn't receive patients when they were admitted to the hospital and they didn't find the medical team when they needed them. Moreover, all studied patients didn't receive hospital orientation, engaged in their treatment plan, and perform any kind of hobbies/leisure activities while they are in the hospital. These results were in concordant with the results of the study done by (Balogun, Agboola, Onigbinde, Ajayi and Iredu, 2018) regarding " Haemophilia Knowledge among Health Care Providers in a Tertiary Hospital Lagos, Nigeria" they revealed lack of adequate knowledge about hemophilia and the management of this rare bleeding disorder among health care providers. Also (Zhao, et al, 2017) pointed out that; hemophilia is a rare disease with complicated diagnosis and management. Patients are helpless because the other personnel are unprofessional.

The study results showed that; all patients did not practice any type of exercises or any type of physiotherapy (therapeutic exercises) because there was not one to help them to practice exercise, and unawareness as well as fear of bleeding was the main reasons of non-practicing therapeutic exercises. The finding agrees with (El Dakhakhny, Hesham, Hassan, El Awady, and Hanfy, 2014) who carried study to determine the impact of health instructions on improving knowledge and practices of hemophilia A adolescents and reported that; all the studied patients didn't practice therapeutic exercises. Also they added that;

physiotherapy, physical activity and sport are basic elements to improve the quality of life and the physical condition, increase strength and resistance, reduce the risk of musculoskeletal lesions and prevent hemophilic atrophy.

Concerning patients' satisfaction more than ninety five percent of patients under this study didn't satisfied with the medical services provided to them, in this line (Rodriguez-Merchan, 2019) reported that; patient satisfaction in hemophilia, a fundamental component of medical care and an indicator of the quality of medical care provided. Moreover, the value of patient satisfaction is particularly apparent in the setting of chronic disease where medical care utilization is high, compliance with therapy is critical and the patient-provider relationship is often long-term.

Regarding to patients' suggestions to improve medical services forty percent of them suggested that; raising health team members and community members' awareness about the disease. These results were in concordant with that of (Balogun, Agboola, Onigbinde, Ajayi and Iredu, 2018) who reported that; the knowledge of the clinical features and management of hemophilia needs to be improved. Lack of adequate knowledge about hemophilia and the management of this rare bleeding disorder among health care providers is of a serious public health concern and likely to increase morbidity and mortality.

As well as, the study of (Nilson, &Schachter, 2011) who stated that the participants' attitudes towards their hemophilia, previous injuries, perceived barriers to seeking treatment, as well as their decision-making process when self-assessing injury. The interviews demonstrated that; communication between the young adults and the health care team was not optimal, with common reference to the ineffectiveness of lecture style education. Gaps in knowledge also emerged regarding bleed identification and management.

On the other hand, limited therapeutic resources in developing countries and ineffectiveness of the existing treatments (increased inhibitor antibodies in some patients) necessitate development of new efficient methods for reducing the disease complications.In the same text (Lazure, et al, 2019) carried study aimed to assess education needs of nurses in thrombosis and hemostasis, and reported that; the need for a more integrated, multidisciplinary approach to care for individuals with bleeding disorders has been highlighted in recent years. Evidence-based education adapted to nurses' needs is essential for a successful evolution.

Also (Valizadeh et al, 2015) pointed that; educating health team members (physicians and nurses) in preventing complications of hemophilia will not only have remarkable economic benefits, but will also inhibit the unexpected complications due to the use of therapeutic products and enhance the patients' quality of life.Hemophilia associations should be recommended for educational programs for patients and caregivers, as well as publishing simple books or brochures in each local language to improve the knowledge (Karimi,Zarei,Haghpanah& Zahedi , 2016).

More than third of studied patients suggested establishing specialized hospitals to provide comprehensive care for patients with hemophilia**.**Infact**,**health insurance opens the first specialized center in Egypt for the treatment and care of hemophilia patients in 18-9-2018. Hemophilia represents a high-cost, low-volume disease hence, in developing countries where ninety percent of hemophilia patients of the world live; it puts the government of the day in a conundrum, i.e. the limited resources of that country earmarked for health need to be distributed amongst management and prevention of various diseases which optimally helps the population of the country. Hence, there is a need for developing research protocols suitable for that country in an evidence-based manner (Ghosh & Shukla 2017).

III. Conclusion

Patients with hemophilia have physical, psychological and social needs. A comprehensive multidisciplinary assessment of the physical, emotional, and social needs of adult hemophilic patients is essential. Clinical pathway may provide comprehensive health care for patients with hemophilia based on patients 'needs to ensure that all important issues are involved, also clinical pathway has been recommended to improve clinical outcomes, decrease mortality rates, and improve cost-effectiveness in health care settings.

IV. Recommendations

Based on the findings of this research, the following recommendations were emerged

- Patients should adhere to comprehensive instructions about hemophilia from health team members.
- Nurse's role as an educator and caregiver for patients with hemophilia should be emphasized.
- All health care professions working hematology units should attend regular in-service training/workshops events to keep abreasted with the latest trends in the care of patients with hemophilia.
- Psychological consultation should be facilitated from specialists for all patients with hemophilia in regular time.
- Awareness of health care professionals should be raised concerning the application of clinical pathway for patients with hemophilia.

- Recreational activities such as (video games / electronic group) should be provided to fill patients' hospital time to decrease feeling of loneliness or depression.
- Availability of coagulation factors as a treatment should be recommended.
- Establish specialized hospitals able to provide care for patients with hemophilia.
- Encourage nurses to apply different types of nursing researches to validate evidence based information in this field.
- Replication of the study on a larger sample and on different geographical settings to allow generalization of findings.

Suggested clinical pathway for patients with all types of hemophilia

This therapeutic clinical pathway is designed to provide comprehensive care based on evidence based practice to improve quality of care, decrease cost, support effective discharge plan, and decrease mortality and morbidity rate for the patients with different types of hemophilia.

Responsible person (leader)

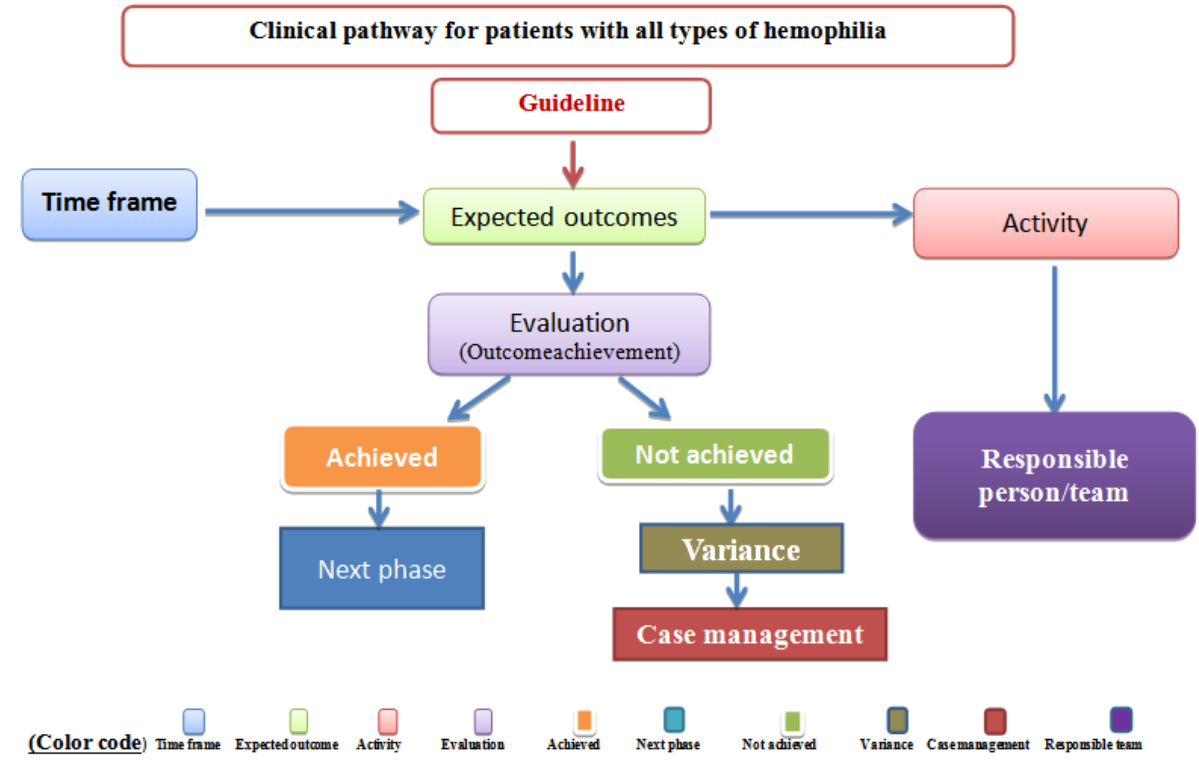
Hematology nurse is the responsible one to coordinate and follow application of clinical pathway for patients with all types of hemophilia because nurse is able to provide comprehensive care for those patients and able to coordinate care and communicate with all members in health team.

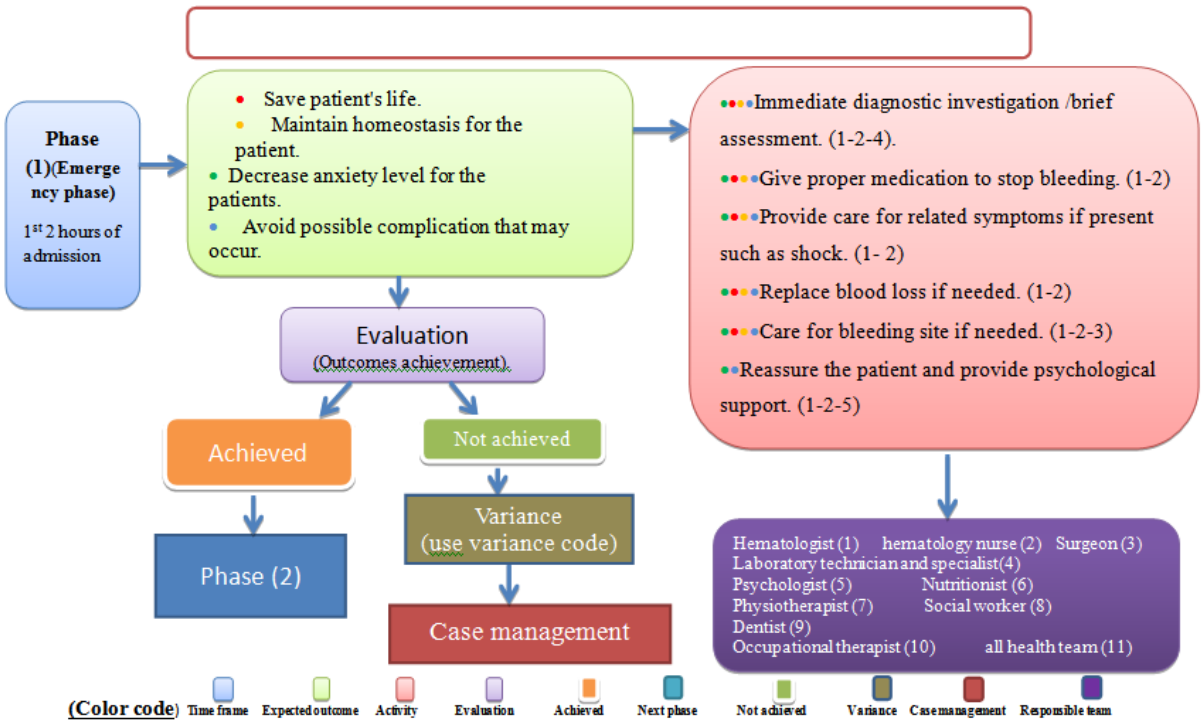
Inclusion criteria

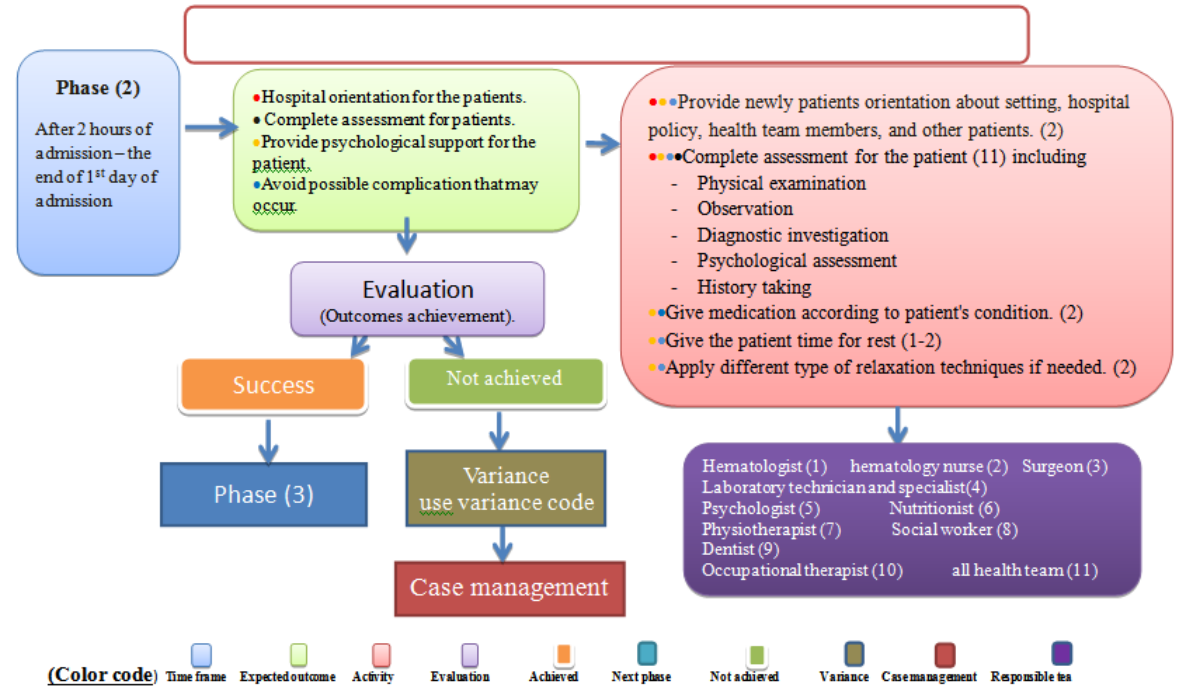
An adult conscious male patient, age from 18-60 years, with all types of hemophilia.

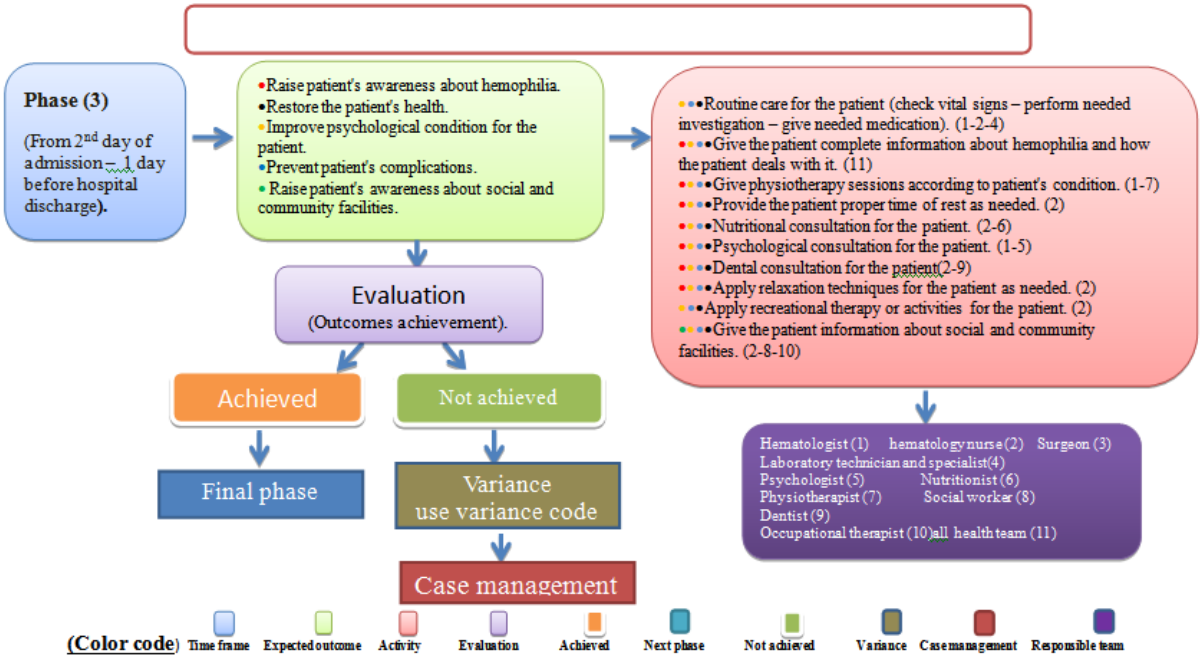
Instructions to use clinical pathway for the patients with all types of hemophilia

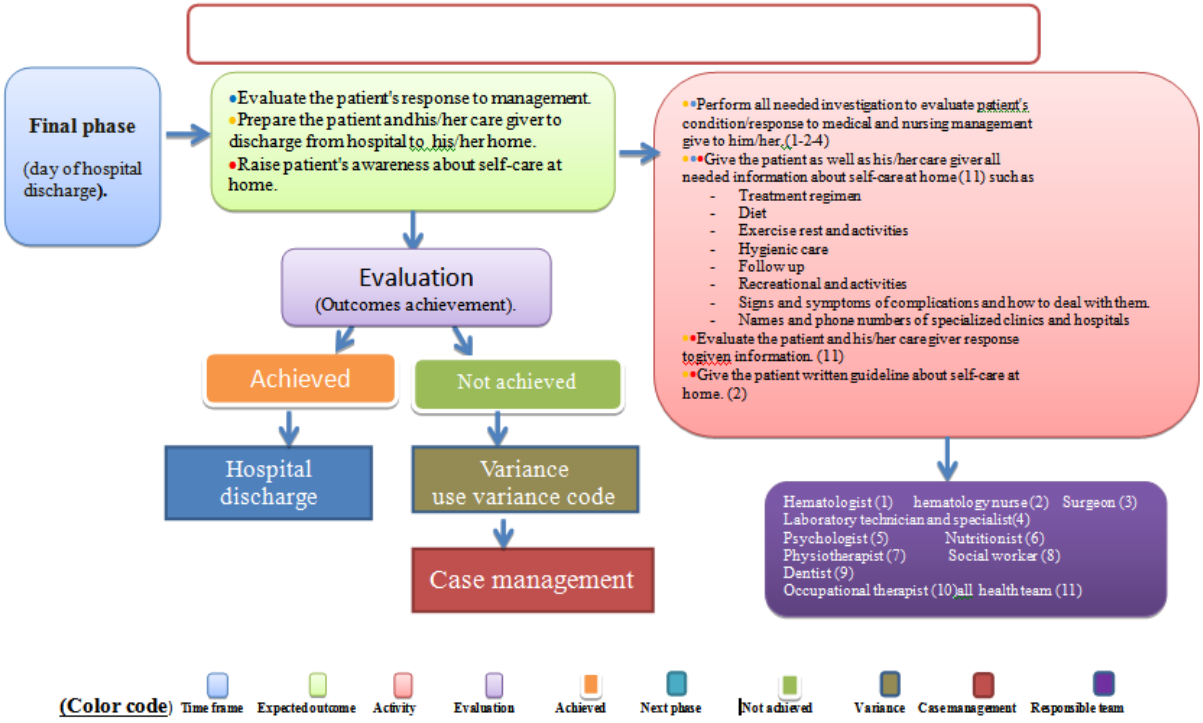
- This pathway is used for patients with all types of hemophilia with inclusion criteria.
- Clinical pathway diagram is color coded; each block indicates specific part in the pathway.
- Expected outcomes are match with activity through color cod in a small circle before each statement.
- Responsible hematology health team is match with activity through assigning numbers presence after each statement.
- Time line for this pathway is from patient's hospital admission until discharge including emergency time.


Clinical pathway for patients with all types of hemophilia
Guideline
Time frame
Expected outcomes
Activity
Evaluation
(Outcomeachievement)
Achieved
Not achieved
Responsible person/team
Next phase
Variance
Case management
(Color code)
Time frame
Expected outcome
Activity
Evaluation
Achieved
Next phase
Not achieved
Variance
Case management
Responsible team


Phase (1)(Emergency phase)
1st 2 hours of admission
Save patient's life.
Maintain homeostasis for the patient.
Decrease anxiety level for the patients.
Avoid possible complication that may occur.
Immediate diagnostic investigation /brief assessment. (1-2-4).
Give proper medication to stop bleeding. (1-2)
Provide care for related symptoms if present such as shock. (1- 2)
Replace blood loss if needed. (1-2)
Care for bleeding site if needed. (1-2-3)
Reassure the patient and provide psychological support. (1-2-5)
Evaluation
(Outcomes achievement)
Achieved
Not achieved
Phase (2)
Variance
(use variance code)
Case management
Hematologist (1) hematology nurse (2) Surgeon (3)
Laboratory technician and specialist(4)
Psychologist (5) Nutritionist (6)
Physiotherapist (7) Social worker (8)
Dentist (9)
Occupational therapist (10) all health team (11)
(Color code)
Time frame
Expected outcome
Activity
Evaluation
Achieved
Next phase
Not achieved
Variance
Case management
Responsible team


Phase (2)
After 2 hours of admission – the end of 1st day of admission
•Hospital orientation for the patients.
• Complete assessment for patients.
•Provide psychological support for the patient.
•Avoid possible complication that may occur.
Evaluation
(Outcomes achievement).
Success
Not achieved
Phase (3)
Variance
use variance code
Case management
•••Provide newly patients orientation about setting, hospital policy, health team members, and other patients. (2)
•••Complete assessment for the patient (11) including
- Physical examination
- Observation
- Diagnostic investigation
- Psychological assessment
- History taking
••Give medication according to patient's condition. (2)
••Give the patient time for rest (1-2)
••Apply different type of relaxation techniques if needed. (2)
Hematologist (1) hematology nurse (2) Surgeon (3)
Laboratory technician and specialist(4)
Psychologist (5) Nutritionist (6)
Physiotherapist (7) Social worker (8)
Dentist (9)
Occupational therapist (10) all health team (11)
(Color code) Time frame Expected outcome Activity Evaluation Achieved Next phase Not achieved Variance Case management Responsible tea


Phase (3)
(From 2nd day of admission – 1 day before hospital discharge).
•Raise patient's awareness about hemophilia.
•Restore the patient's health.
•Improve psychological condition for the patient.
•Prevent patient's complications.
• Raise patient's awareness about social and community facilities.
Evaluation
(Outcomes achievement).
Achieved
Not achieved
Final phase
Variance
use variance code
Case management
•••Routine care for the patient (check vital signs – perform needed investigation – give needed medication). (1-2-4)
•••Give the patient complete information about hemophilia and how the patient deals with it. (11)
•••Give physiotherapy sessions according to patient's condition. (1-7)
•••Provide the patient proper time of rest as needed. (2)
•••Nutritional consultation for the patient. (2-6)
•••Psychological consultation for the patient. (1-5)
•••Dental consultation for the patient(2-9)
•••Apply relaxation techniques for the patient as needed. (2)
••Apply recreational therapy or activities for the patient. (2)
•••Give the patient information about social and community facilities. (2-8-10)
Hematologist (1) hematology nurse (2) Surgeon (3)
Laboratory technician and specialist(4)
Psychologist (5) Nutritionist (6)
Physiotherapist (7) Social worker (8)
Dentist (9)
Occupational therapist (10) all health team (11)
(Color code) Time frame Expected outcome Activity Evaluation Achieved Next phase Not achieved Variance Case management Responsible team



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