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PREMARITAL COUNSELING CLINICAL TESTS GUIDELINES

IN PRIMARY HEALTH CARE CENTERS IN IRAQ

DISCLAIMER

This guideline has made possible through support provided by the U.S. Agency for International Development (USAID) under Primary Health Care Project in Iraq (PHCPI) implemented by University Research Co., LLC. This guideline has been developed in Iraq in close collaboration with the Ministry of Health (MoH) in February 2013

List of Contents	
List of content	2
List of figures, and boxes	3
List of abbreviation	4
Chapter One: Introduction	5
Objectives	6
Chapter Two: Plan of action in premarital counseling	7
Premarital counseling risk assessment form	7
Premarital counseling clinical pathway	7
Chapter Three: Premarital guidelines for most common heamoglobinopathies in Iraq	11
Overview	11
Beta thalassemia	12
Sickle cell disorder	17
Chapter Four: Premarital guidelines for sexually transmitted diseases	20
Hepatitis B	20
Syphilis	21
Acquired immunodeficiency syndrome	23
Annex One: Premarital counseling risk assessment form	25
Annex Two: Performance Checklist	26
Annex Three: Preconception counseling	28
Annex Four: Appropriate age for marriage of women	30
Annex Five: Consanguineous marriages (endogamy)	31
References	32

Figures and Boxes	
Box 1: Main objectives of premarital counseling services	6
Box 2: Premarital counseling in special consultations	10
Box 3: Indications for referral to genetic department	10
Figure 1: Premarital counseling protocol for β -thalassemia	16
Box 4: Indications for sickle cell testing in premarital counseling	19
Box 5: Causes of false VDRL test results	22
Figure 2: Premarital counseling protocol for syphilis	24

Abbreviations

B-thal	Beta-thalassemia
G6PD	Glucose 6 Phosphate Dehydrogenase
Hb	Hemoglobin
HBB	Beta Globin
HBIG	Hepatitis B Immune Globulin
HBsAg	Hepatitis B Surface Antigen
HBV	Hepatitis B Virus
HCP	Health Care Provide
HIV	Human Immunodeficiency Virus
HPLC	High Performance Liquid Chromatography
MCH	Mean Corpuscular Hemoglobin
MCV	Mean Corpuscular Volume
PC	Premarital Counseling
PEP	Post-exposure Prophylaxis
PHC	Primary Health Care
SCD	Sickle Cell Disease
STD	Sexually Transmitted Disease
TPHA	Treponema Pallidum Heamagglutination
VDRL	Venereal Disease Research Laboratory

Premarital Counseling Clinical Guidelines

Chapter 1: Introduction

Premarital counseling (PC) is a method of advice and guidance for those intending to marry to enhance their understanding and selection and to improve their quality of life.

This service depends on communication and persuasion skills to enhance the ability to choose after obtaining the information necessary and make inquiries without coercion since people who want to marry are the decision-makers.

The aim of premarital counseling is to provide baseline assessment of would-be married couples; raise their level of health; and to identify and reduce the reproductive genetic risks.

Premarital counseling started in Iraq from long period of time. It has been carried on in many specialized centers distributed throughout the country. The current plan of Ministry of Health is the incorporation of this service as a routine ongoing process at the primary health care level.

Premarital Counseling is provided in health centers as part of the primary health care preventive measures based on the protocols and guidelines laid down by the Ministry of Health. This service is provided by physicians, and supported by trained paramedics or other health care providers (HCPs).

Premarital Counseling will identify couples at high risk, by subjecting all couples intending to marry for screening by history taking, physical examination and laboratory investigations.

Accordingly, advice is provided in the form of further investigation or referral to secondary level if needed, treatment advice, health education and promotion, and counseling regarding their health status.

The following pages represent the first trail implemented in Iraq in order to provide guidelines that organize the work in premarital counseling services. This can be achieved by following standardized specific roles and recommendations for the process of counseling at the different Health facilities.

Objectives

The main objectives of premarital counseling have been summarized in the following box:

Box 1: Main Objectives of Premarital Counseling Services

1. To reduce the incidence of common haemoglobinopathies in Iraq, e.g. thalassemias and sickle cell anemia.
2. To reduce other hereditary disorders by identifying problems followed by counseling.
3. Counseling regarding high-risk behaviors, including those related to HIV, Hepatitis B, and other infectious diseases.
4. Early detection and treatment of some sexually transmitted diseases.
5. To promote awareness regarding reproductive health, family planning, and healthy lifestyles.
6. To provide couples with medical, social, and psychological support.
7. To provide immunizations as required.

Counseling should be provided to the couples together, unless a specific problem requires personal confrontation. Couples should understand that counseling **does not** guarantee the absence of congenital anomalies, nor does it assess their fertility.

The certificate on premarital counseling risk assessment form will be issued only after counseling both partners together, stating that “couples were counseled together, and the decision of marriage is left to couples themselves”. The certificate will be registered, and given a registration number and duly signed and stamped by the examining physician and the director of health facility.

The following pages are the guidelines for all care givers to help them in dealing with couples who want to marry. They assist in detecting the majority of common haemoglobinopathies and sexually transmitted diseases in each partner. They provide also certain clues to determine the possibility of transmission of inherited diseases to offspring.

Chapter 2: Plan of Action in Premarital Counseling

Premarital Counseling Risk Assessment Form

The assessment form in premarital examination (Annex 1) should include the following:

1. Administrative information: name of health institution, sequential number of form, number of certificate and its date of issue.
2. Socio-demographic data: name, age, and occupation, of partners; in addition to the degree of consanguinity between them.
3. Family history: inherited blood diseases (thalassemia, sickle cell anemia, and hemophilia), mental disability, and congenital physical handicap. Any important notes should be recorded.
4. Laboratory investigations: blood grouping and Rh factor, RBC indices (MCV and MCH), VDRL, HBsAg, and tests for HIV and tuberculosis.
5. Procedures: the female partner should be given a dose of tetanus toxoid after checking her immunization status. The field concerning this action is labeled.
6. Signatures of couple, names and signature of examining doctor and director, and stamp of health institution.

Premarital Counseling Clinical Pathway

- Every couple who want to get married should be subjected to Premarital Counseling.
- Premarital Counseling is provided by physicians with the help of trained paramedic or other health care provider.
- Couples are referred to health center within the catchment area (one of the health centers they belong to) for getting the service of premarital counseling.
- Appointments are arranged by the relevant clerk or health care provider
- Usually Premarital Counseling is completed in 2-3 visits in normal situation.

I. The First Visit (to the Paramedics/HCP)

1. Basic data and address on the PC Risk Assessment Form is filled in, either by the HCP or the client.
2. The degree of consanguinity between the couple must be identified.
3. HCP should take family history as per the PC Risk Assessment Form
4. When applicable, check the weight and height and calculate the body mass index (BMI).
5. Request blood sample from both partners for required lab investigations and record the results when coming back.
6. The female partner should be given a dose of tetanus toxoid, after checking history of tetanus vaccination, and the field concerning this action on the Premarital Counseling Form is labeled.
7. Premarital educational booklet will be given to each client. The booklet includes information on the following:
 - The concept and aim of Premarital Counseling
 - Common haemoglobinopathies in Iraq
 - Sexually transmitted diseases (STI)

- Healthy life-style
 - Concept of family planning and methods of contraception.
 - Breast self-examination, the maneuver and frequency
8. Each partner should sign the premarital counseling consent.
 9. The phone number and any possible way for contact with the partners should be requested.
 10. Each client should be given an appointment for counseling before they leave.

II. Preparation for the next visit

1. Before the next appointment (to the doctor), the Health Care Provider has to review the contents of Premarital Counseling Form, i.e. Family history and Lab reports; if any doubt should discuss it with the physician.
2. If any abnormal result, such as positive Hepatitis B virus, Syphilis etc., a separate appointment for each partner should be arranged.

III. The Second Visit (to the Physician)

1. When possible, the initial interview with each partner should be held individually and separately
2. Review the Premarital Counseling Risk Assessment Form,
3. Take the clinical history, concentrate on the following:
 - A. Medical/Surgical/Psychological History
 - a. Significant systemic illness such as: Hypertension, Diabetes mellitus, Heart problems, Epilepsy etc.
 - b. Previous surgical history
 - c. History of mental illness
 - d. History of sexually transmitted diseases (STI) such as: syphilis, gonorrhea, hepatitis B, genital warts and ulcer, and urethral/vaginal discharge.
 - e. History of blood transfusion (date, frequency, place etc.)
 - f. History of current medications and allergy to certain drugs
 - g. Family history of hereditary diseases or genetic problems
 - Sickle cell, thalassemia, hemophilia, G6PD deficiency, or frequent blood transfusion
 - Congenital anomalies in the family
 - Mental retardation
 - Family history of hypertension, diabetes, etc.
 - h. If previously married, history of baby with congenital abnormality.
 - B. Habits and Risk behavior such as:
 - Smoking, Alcohol consumption, substance abuse
 - C. Others, such as:
 - Tattooing, and multiple ear piercing
 - Practiced some religious ceremonies involving sharp objects and bloodletting (Hejamah).
4. Perform physical examination, concentrate on:
 - A. General Physical Examination:

- General look of patient and signs of anemia, jaundice and cyanosis.
- B. Systematic physical examination:
- Neck for thyroid, Chest, CVS and Abdomen.
- C. Vital signs
- Blood pressure and pulse
5. Review lab reports, referring accordingly to:
 - Premarital Guidelines for Most Common Hemoglobinopathies in Iraq.
 - Premarital Guidelines for Sexually Transmitted Diseases.
 7. Assess the condition of each partner
 8. When applicable and needed, do counseling for each partner separately at first.
 9. Final counseling should be offered in the presence of both partners.
 10. Issue the certificate on PC Risk Assessment Form.
 11. In case of abnormal findings or results, special actions and interventions should be undertaken (box 2 and 3).

IV. Documentation

- In order to avoid the loss of lab tests and make benefit from them in the future, it is advised to undertake the following:
 - A copy from the Risk Assessment Forms will be kept in a Box File till the couple opens a family folder in a health center, and then it will be transferred to their HC and kept in their Family Folder.
 - personal card; or even a copy of Risk Assessment Forum, is given to each client with his/her laboratory results

Box 2: Premarital Counseling in Special Consultations

- Partner with abnormal finding/s or result/s, which needs further assessment, evaluation, management and counseling will be offered treatment locally or referred to secondary care.
- Extra appointment/s can be arranged if necessary.
- Couples found to have a risk of an affected offspring will be referred for genetic counseling at the secondary care.
- Both partners should be advised to share data by themselves.
- If they agree, an appointment is booked for counseling both partners together.
- In the cases required referral to secondary care, issue the Premarital Counseling Certificate based on a written feedback from the secondary care.
- When there is a documented reproductive genetic risks from this marriage, it is advised that the doctor to write “from medical aspect, this marriage may lead to the following disease in the next generation:”
- The partner of person with sexually transmitted disease should be notified with registration of actions and outcomes.

Box 3: Indications for Referral to Genetic Department

I. Couples at High Risk

1. Both partners are carrier of sickle cell disease (sickle cell trait).
2. Both partners are carrier of B-thalassemia (B-thal trait).
3. Each partner has different abnormal Hb, e.g. one has sickle cell trait and the other has B-thal trait.
3. Blood results need clarification.
4. Family history of other genetic diseases.
5. Family history of congenital or chromosomal abnormalities.

II. Couples Who May Be at Risk

One partner has sickle cell trait or B-thal trait and the other partner has hypochromia microcytosis and/or HBA2 between 3.4-3.7

Note: If you are sure of the diagnosis, no need to refer the couples when both partners are having alpha-thalassemia gene.

Chapter 3: Premarital Guidelines for Most Common Hemoglobinopathies in Iraq

Overview

With approximately 7% of the worldwide population being carriers, hemoglobinopathies are the most common monogenic diseases and one of the world's major health problems. They were originally found mainly in the Mediterranean area and large parts of Asia and Africa.

International migration has spread them from those areas all over the world. In many parts of world today, hemoglobin (Hb) defects are classified as endemic diseases.

Iraq is one of the countries which are endemic with different types of hemoglobinopathies.

There are several epidemiological studies on their frequency. The prevalence of gene carriers is high giving a figure of millions among the entire population. The total number of patients diagnosed with these diseases in the MoH laboratories during the last years is tens of thousands. Thus, detection and advice about these disease becomes the corner stone element in Premarital Counseling in order reduce the burden of this problem.

Types and Composition of Adult Hemoglobin

Normal hemoglobin (Hb) in adults is composed of the following:

1. Hb A: this is composed of two alpha and two beta chains, it constitute 95% of total hemoglobin.
2. Hb A2: this is composed of two alpha and two delta chains, it constitute 3% of total hemoglobin.
3. Hb F (fetal): this is composed of two alpha and two gamma chains, it constitute 2% of total hemoglobin.

Basic types of hemoglobinopathy

The umbrella term "hemoglobinopathy" includes all genetic hemoglobin disorders. These are divided into two main groups. Both are caused by mutations and/or deletions in the α - or β -globin genes of hemoglobin.

- **Thalassemia syndromes:** These are autosomal recessive conditions resulting from the decreased or absent synthesis of globin chains. Alpha and beta thalassemia are the result of deficient or absent synthesis of α and β chains, respectively. Hemoglobin structure in these cases is normal.
- **Abnormal hemoglobins (structural hemoglobin variants):** This group of autosomal dominant inherited hemoglobin disorders is caused by structural defects in hemoglobin synthesis resulting from an altered amino acid sequence in the α - or β - chains.

The most common types of hemoglobinopathies enrolled in Iraqi Premarital Counseling are discussed below.

I. Beta-thalassemias

Beta thalassemia (β -thal) syndromes are the result of insufficient (β^+) or absent (β^0) production of β -globin chains. Their molecular causes are β -globin gene mutations.

β -thalassemia is prevalent in populations in the Mediterranean, Middle East (including Iraq), Transcaucasia, Central Asia, Indian subcontinent, and Far East. It is also common in populations of African heritage. Hematological changes become manifest from between the ages of three months and six months onwards.

Genetic issues:

- Risk of being born with thalassemia and the severity of disease is dependent on the genetic composition of parents.
- The gene for β -thalassemia is autosomal recessive. However, a person with even one of these genes (heterozygous, carrier state) will have some hemoglobin changes, although these changes are often minor.
- The transmission of defective gene and risk of being affected are outlined as:
 - One parent with thalassemia minor (heterozygous) and the other is normal – 50% chance of each child having thalassemia minor and 50% to be normal.
 - Both parents with thalassemia minor (heterozygous) – 25% chance of each child having thalassemia major (homozygous), and 50% chance of each child also having thalassemia minor and only 25% chance that the child will not have some form of thalassemia.

Clinical picture of β -thalassemia

- Thalassemia Minor (heterozygous – carrier state)
 - Patient is a silent carrier of the thalassemia gene and there are no clinical symptoms.
 - Patients may have mild anemia, mild microcytosis that does not interfere with normal daily function.
 - However, mild anemia may become worse during times of physiologic stress – pregnancy, co-existing iron deficiency, chronic illness, old age.
- Thalassemia Major (homozygous)
 - These patients are the most symptomatic and ill.
 - Signs and symptoms in untreated children usually appear in early childhood, such as:
 - Severe anemia – microcytic, hypochromic.
 - Increased erythropoiesis (RBC production), which causes an expansion in bone marrow tissue.
 - Thinning of bones caused by increased bone marrow production, with osteoporosis and osteomalacia – especially seen in face and skull.
 - Splenomegaly caused by increased hemolysis and destruction of abnormal RBC.
 - Increase iron absorption which can lead to iron overload in tissues.

➤ **If untreated, patients often have the following symptoms:**

- Pale or jaundiced appearance due to anemia and hemolysis.
- Fatigue and lack of energy.
- Abnormal growth patterns and delayed puberty
- Pathologic fractures of long bones.
- Heart failure due to anemia and iron deposition in heart muscle.
- Enlarged spleen, leading to cirrhosis of the liver.
- Frequent ulcers on the legs and gall bladder disease.

Note: There is certain type of thalassemia called thalassemia intermedia in which the clinical severity of the disease is somewhere between the mild symptoms of the β thalassemia trait and the severe manifestations of β thalassemia major.

Thalassemia intermedia is inherited and may result from a wide variety of genotypes. Certain homozygous β thalassemia alleles have produced this condition. Additionally several forms of combined heterozygous thalassemia can also result in a clinical course consistent with thalassemia intermedia.

Laboratory Diagnosis of Thalassemia:

- MCV and MCH decreased – RBC indices are decreased, even in thalassemia minor – This is the first screening evidence of possible thalassemia (or other form of anemia).
- Mild to severe anemia (depending on severity of thalassemia):
 - HbA2 increased to >3.5%
 - HbF increased, sometimes to >90%
- Peripheral blood smear shows abnormal RBC – microcytic, tear-drop shapes, and target cells.
- Serum iron and ferritin are usually increased.
- May have increased bilirubin, and abnormal liver function studies.

Treatment Options for Beta - Thalassemia

I. For Thalassemia Minor:

- Usually no treatment required.
- May need occasional transfusion during times of physiologic stress.
- Supplementation with folic acid often recommended.

II. For Thalassemia Major:

- Most patients require periodic transfusion to maintain adequate functioning hemoglobin and suppress increased erythropoiesis.
- With transfusions and maintenance of Hb. Level at 10-11 mg/dl. complications such as hypersplenism, fractures, infections, and orthopedic complications are less common.
- Periodic transfusions lead to further iron overload, and need for iron chelation therapy with deferoxamine.
- Should supplement with folic acid because of increased erythropoiesis (production of RBC).
- Radical treatments include bone marrow transplantation, or experimental gene therapy.

- Early management of complications.

Prevention strategies

- The only prevention of thalassemia major is to prevent passing down the genes to a child.
- Primary strategy in Middle East is counseling of couple when both male and female found to be heterozygous thalassemia carriers.
- In some countries, prenatal diagnosis can be made on fetus of two thalassemia carriers (small sample of fetal blood, or genetic analysis) to determine if fetus is affected.

Premarital Screening for thalassemia:

- All couples to be married will need certificate of probable thalassemia status.
- Both man and woman should be screened; although only one can be screened, and if positive, then request screening of second.
- Screening test – RBC cell size and hemoglobin concentration (MCV and MCH) – should be greater than 80 and 27, respectively
- If both RBC indices for one member of couple are normal (even if second member has abnormal low results), certificate can be given.
- If at least one of RBC indices on BOTH members of couple has abnormal low value, BOTH patients referred for hemoglobin electrophoresis (or HPLC) as definitive test.
- Other findings, such as blood smear findings, Hb level, iron or ferritin levels...etc, can provide additional information, but not as accurate as hemoglobin electrophoresis.

Explanation of Results of Screening and Confirmatory Tests

A. For Screening Test:

- If screening test negative – reassure patient and provide appropriate signed statement.
- If screening test positive – give further counseling:
 - Explain need for further testing – hemoglobin electrophoresis or HPLC and possibly iron testing – as specific test to determine cause of abnormal results and thalassemia status.
 - Arrange for hemoglobin electrophoresis/HPLC test.
 - Answer patient questions about testing and thalassemia.

B. For Confirmatory Tests:

- If hemoglobin electrophoresis test/HPLC positive for thalassemia carrier state (Hb.A2 > 3.5% and/or Hb.F increased) – refer patient to specialized Thalassemia Counselor for further counseling.
- If hemoglobin electrophoresis tests normal, continue testing to determine cause of microcytosis and/or hypochromia.
 - Most common cause is iron deficiency anemia, so request serum iron and ferritin, and total iron binding capacity (TIBC) levels.
 - If iron deficiency confirmed by low iron and ferritin level or high TIBC, treat with iron supplement and follow-up.

- If iron deficiency not confirmed (iron tests normal), patient may have a silent thalassemia trait or other problem, and should be referred to appropriate specialist.

NOTE: Alpha thalassemia occurs more frequently in South-East Asia and is not screened routinely during premarital counseling.

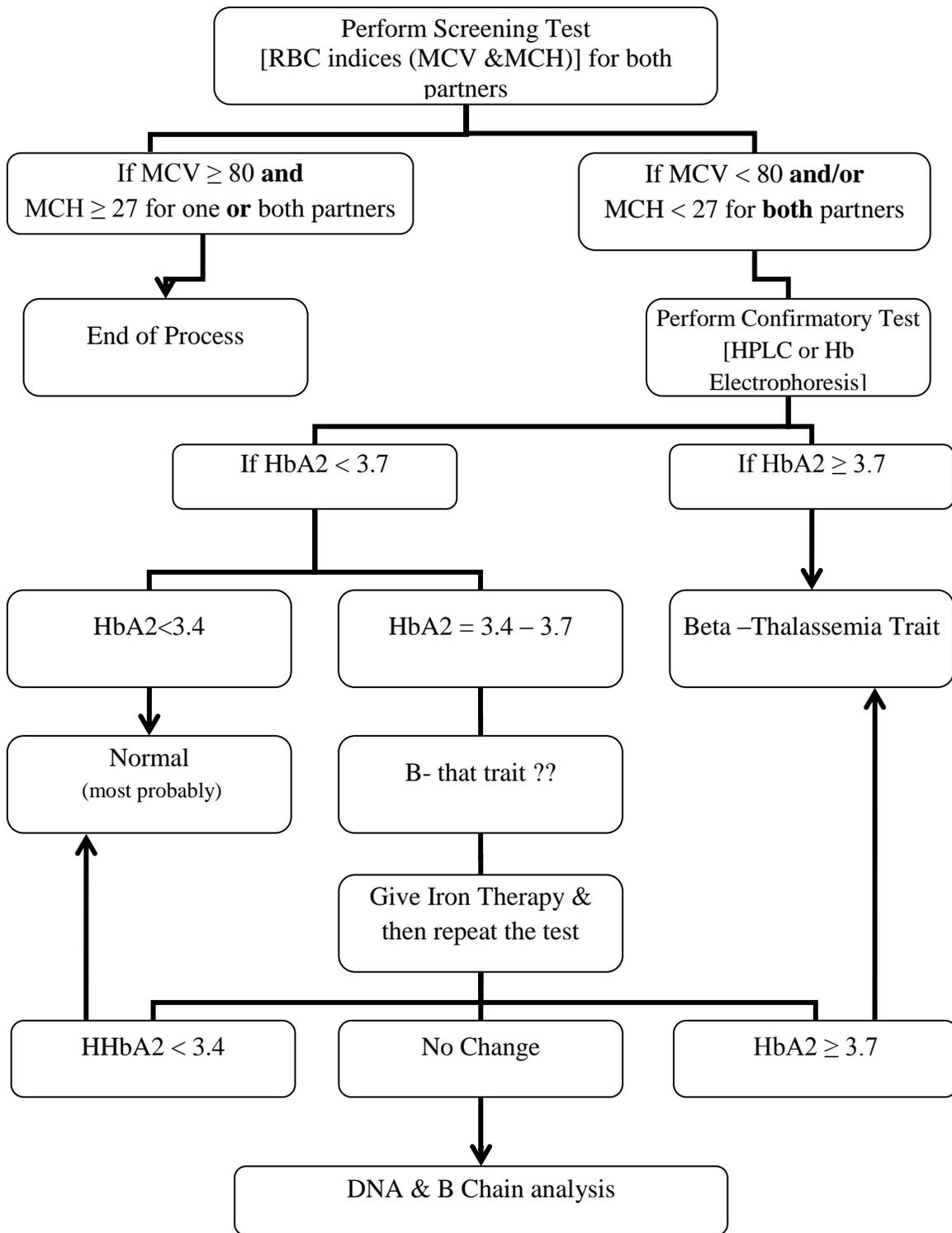


Figure 1: Premarital Counseling Protocol for β -Thalassemia

II. Sickle Cell Disorder

The term “sickle-cell disorder” includes all manifestations of abnormal hemoglobin S levels (proportion of HbS >50%). These include spectrum ranging from homozygous sickle-cell disease (HbSS) through mixed heterozygous hemoglobinopathies (HbS/β-thalassemia, HbSC disease, and other combinations) to purely heterozygous sickle cell trait (HbSA).

Sickle cell disease is most common in people living in or originating from sub-Saharan Africa. The disorder also affects people of Mediterranean, Caribbean, Middle-Eastern (including Iraq), and Asian origin.

Genetic issues:

- Sickle cell disease is inherited in an autosomal recessive manner.
- If both parents are carriers of beta-globin (HBB) mutation, each offspring has at conception a 25% chance of being affected with the disease, a 50% chance of being an asymptomatic carrier, and a 25% chance of being unaffected and not a carrier.
- If one parent is homozygous and the other parent is heterozygous for an HBB mutation, each offspring has a 50% chance of being affected and a 50% chance of being an asymptomatic carrier.
- If both parents are homozygous, all offspring will be affected.

Clinical picture of sickle cell disorder

- Sickle cell trait (heterozygous – carrier state)
 - These individuals do not express symptoms of sickle cell disease
 - Individuals normally do not have anemia, except in the cases of mixed heterozygous hemoglobinopathies (e.g. HbS/β-thalassemia, HbSC disease, and other combinations).
 - Although, Heterozygotes (carriers) are generally asymptomatic, but they may develop complications under extremes of physical exertion, dehydration, and/or altitude.
- Sickle cell disease (homozygous – disease state)
 - Most individuals with sickle cell disease are healthy at birth and become symptomatic later on, after fetal hemoglobin (Hb F) levels decrease and hemoglobin S (Hb S) levels increase.
 - The severity of disease manifestations varies from severe to minimal, even in individuals with the same HBB mutation status.
 - The main causes of death are infection, acute chest syndrome, pulmonary artery hypertension, and cerebrovascular events.
 - **If untreated, patients often have the following symptoms:**
 - Vaso-occlusive events result in tissue ischemia leading to acute and chronic pain as well as organ damage that can affect any organ in the body, including the bones, lungs, liver, kidneys, brain, eyes, and joints.
 - Dactylitis (pain and/or swelling of the hands or feet) in infants and young children is often the earliest manifestation of sickle cell disease.

- In children the spleen can become engorged with blood cells in a “splenic sequestration crisis.” The spleen is also particularly subject to infarction and the majority of individuals with SCD are functionally asplenic in early childhood, increasing their risk for certain types of bacterial infections.
- Chronic hemolysis results in varying degrees of anemia, jaundice, cholelithiasis, and delayed growth and sexual maturation. Individuals with the highest rates of hemolysis are predisposed to pulmonary artery hypertension, priapism, and leg ulcers but are relatively protected from vaso-occlusive pain.

Laboratory Diagnosis of Sickle Cell Disease:

- The presence of significant quantities of Hb S by high-performance liquid chromatography (HPLC), isoelectric focusing, or (less commonly) cellulose acetate or citrate agar electrophoresis
- The lack of a normal β -globin gene on Molecular Genetic Testing.
- Peripheral blood smear shows Sickle cells, nucleated red blood cells, and target cells may be seen.
- MCV and HbA2 may be normal or reduced depending on specific genotype of disorder.

Treatment for Sickle Cell Disorder:

I. For Sickle Cell Trait:

- People with sickle cell trait can live full lives and enjoy most of the activities that other people do.
- There are things that those people can do to stay as healthy as possible. Below are a few examples:
 - *Get regular checkups.* Regular health checkups with a primary care doctor can help prevent some serious problems.
 - *Prevent infections.* Common illnesses, like the flu, can quickly become dangerous for a child with sickle cell disease. The best defense is to take simple steps to help prevent infections.
 - *Learn healthy habits.* People with sickle cell trait should drink 8 to 10 glasses of water every day and eat healthy food. They also should try not to get too hot, too cold, or too tired.

II. For Sickle Disease:

- The mainstay is good hydration and avoidance of climate extremes, extreme fatigue, and activities leading to inflammation.
- Hydroxyurea can decrease the frequency and severity of vaso-occlusive processes, reduce transfusion needs, and increase life span.
- Chronic red blood cell transfusion is indicated in children with either a history of or risk factors for stroke and other specific complications, such as pulmonary hypertension and chronic renal failure.
- Aggressive education on the management of fevers; prophylactic antibiotics, including penicillin in children; up-to-date immunizations; and iron chelation therapy for those with iron overload.
- Early management of complications.

- Periodic monitoring investigation and follow-up examination

Prevention strategies

- As for thalassemia, the only way for prevention of sickle cell disease is to prevent passing down the genes to a child.
- It is appropriate to provide counseling of couple when both male and female found to be heterozygous sickle cell carriers.
- In some countries, Pre-implantation genetic diagnosis (PGD) may be available for families in which the disease-causing mutations have been identified.

Premarital Screening and counseling for sickle cell disorder:

- Routine screening for sickle cell disorder is not currently available in Iraq. However, this action can be offered to couple if any member of them is at high risk of being carrier (Table 4).
- Those partners should be referred to health facility where appropriate investigations and counseling for sickle cell disorder are available.
- Carrier detection for HBB mutations involving abnormal hemoglobin (e.g. sickling disorder) is most commonly accomplished by HPLC or hemoglobin electrophoresis.

Box 4: Indications for Sickle Cell Testing in Premarital Counseling

1. Positive family history of sickle cell disorder
2. Suggestive personal history of having sickling trait:
 - a. Recurrent urinary tract infections in women or gross hematuria of unclear etiology
 - b. History of splenic infarction at high altitude, with exercise, or with hypoxia
 - c. History of glaucoma or recurrent hyphema following a first episode of hyphema
 - d. History of exercise related life – threatening complications or exertional heat illness (e.g. exertional rhabdomyolysis, heat stroke, or renal failure)
3. The other partner is already known to have sickling disorder, whether trait or disease.

Note: It must be kept in mind that non-sickle β -globin disorders (e.g., β -thalassemia) can interact with the SCD-causing mutation leading to clinically significant disease.

For example, if one parent has sickle cell trait and the other has β -thalassemia trait, it would be correct to state that, although one parent is not a sickle cell carrier, there is still a 25% chance that each pregnancy would have a significant hemoglobinopathy.

Therefore, partners of individuals who are known to carry sickle cell trait should be offered a thalassemia screening panel that includes hemoglobin electrophoresis or HPLC to screen for carrier status for sickle cell trait and other β -globin disorders.

Note: Premarital screening for other variants of structural hemoglobin abnormality (e.g. HbC, and HbE) is not performed routinely during premarital counseling.

Chapter 4: Premarital Guidelines for Sexually Transmitted Diseases

Premarital Counseling visits represent a good opportunity for detection and treatment of sexually transmitted diseases, as well as prevention of their transmission to the partner. It provides also the time for giving the advices and instructions of their risks, modes of transmission, and ways of avoidance and protection from them.

I. Hepatitis B

Hepatitis B is caused by infection with the hepatitis B virus (HBV). The incubation period from the time of exposure to onset of symptoms is 6 weeks to 6 months. HBV is more infectious and relatively more stable in the environment than other blood borne pathogens like HCV and HIV. Viral hepatitis is endemic in Iraq. The prevalence of HBsAg is 2-3% in the normal population. Both couples should do **HBsAg** as a routine giving that positive test will pick up 99% of cases, in addition to being also cheap and easy test.

Explanation of results of HBsAg test

- If Negative HBsAg- Reassure the partner and give the appropriate counseling on STDs
- If **Positive** HBsAg
 - **Refer to secondary care**
 - Before referral, do counseling regarding:
 - Relieve anxiety
 - Risk: HBV infection can be self-limited or chronic. Risk for chronic infection is inversely related to age at acquisition. In adults, only approximately half of newly acquired HBV infections are symptomatic, and approximately 1% of reported cases result in acute liver failure and death. Among persons with chronic HBV infection, the risk for premature death from cirrhosis or hepatocellular carcinoma (HCC) is 15%–25%.
 - Prevention: Two products have been approved for hepatitis B prevention: hepatitis B immune globulin (HBIG) and hepatitis B vaccine.
 - ✓ HBIG provides temporary (i.e. 3–6 months) protection from HBV infection and is typically used as PEP (post-exposure prophylaxis) either as an adjunct to hepatitis B vaccination in previously unvaccinated persons or alone in persons who have not responded to vaccination.
 - ✓ Hepatitis B vaccine provides protection from HBV infection when used for both pre-exposure vaccination and PEP.

Counseling:

- Partner notification is mandatory.
- Vaccination is advised for the partner (three doses at 0 - 1 - 6 months).
- Do (Anti HBsAb) within 1-4 weeks from the third dose to check immunity

- ✓ If (Anti HBsAb) test is **Positive** (Immune) - Give a booster dose every 5 years
- ✓ If (Anti HBsAb) test is **Negative** (Non-immune) - Refer to secondary care

Follow-up:

The following investigations should be requested for the patient every 6 months:

1. Hepatitis B profile (HBsAg, Anti HBc(IgM), HBeAg, AntiHBeAb)
2. Liver function test (LFT), prothrombin time(PT), and complete blood count (CBC).
3. Liver Ultrasound

The result of these investigations will determine the patient status:

- If found **Carrier**
(Anti HBeAb positive, HBeAg negative with normal LFT and Ultrasound)
 - Counseling
 - Repeat check up every 6 months
 - Family screening
- If found **Active**
(HBeAg positive, elevated LFT or abnormal liver ultrasound)
 - Refer to secondary care

II. Syphilis

Syphilis is a systemic disease caused by *Treponema pallidum*. On the basis of clinical findings, the disease has been divided into a series of overlapping stages, which are used to help guide treatment and follow-up.

Stages of Syphilis:

1. Primary stage: there is ulcer or chancre at the infection site.
2. Secondary stage: its manifestations include, but are not limited to, skin rash, mucocutaneous lesions, and lymphadenopathy.
3. The latent stage: this begins when primary and secondary symptoms disappear. Latent syphilis acquired within the preceding year is referred to as early latent syphilis; other cases of latent syphilis are late latent syphilis.
4. Tertiary stage: symptoms include difficulty coordinating muscle movements, paralysis, numbness, gradual blindness, and dementia. The disease damages the internal organs, including the brain, nerves, eyes, heart, blood vessels, liver, bones, and joints. This damage can result in death.

Premarital Screening for Syphilis:

- Do Venereal Disease Research Laboratory (VDRL) test:
 - If the test is negative then the partner most likely to not have the disease
 - If **Positive** (Reactive) refer to secondary care level to do TPHA
- TPHA (Treponema pallidum hemagglutination test):

This test used to confirm the diagnosis of syphilis in patients with positive (reactive) VDRL. It usually remains positive for life. False positive results may occur in 1-2%, e.g. systemic lupus erythematosus and Lyme disease.

 - If TPHA is negative then the partner most likely to not have the disease
 - If TPHA is positive, then management is provided accordingly

Box 5: Causes of False VDRL Test Results

I. False Negative VDRL, decreased sensitivity in 1/3 of patients in early primary and during late syphilis (i.e. VDRL may be non-reactive), so the physician should be alert to these situations

II. False Positive VDRL (usually titer < 1:8 [1:2-1:8]), causes include:

1. Autoimmune disease
2. Connective tissue disease
3. Concomitant viral infection
4. Pregnancy
5. Elderly
6. IV drug abuse
7. Recent immunization
8. Dermatological diseases
9. Febrile illness
10. Multiple blood transfusion

Follow-up

- Repeat clinical examination and serology (VDRL quantitative titer) after 1, 3, 6, 12 and 24 months to assess treatment efficacy and compliance.
- Decline in VDRL titer of > four-folds indicates cure (i.e., compared with the maximum or baseline titer at the time of treatment)
- VDRL titer should become at least 4 times lower within 6 months after treatment of primary or secondary syphilis, and within 12 to 24 months after treatment of latent or late infection.
- Patients with clinical manifestations that persist or recur or have a sustained four-fold increase in VDRL test titer probably failed treatment or were reinfected.

Counseling

- All patients with syphilis should be counseled for partner notification, health education, and confirmation of any past treatment history.
- For patients with primary syphilis, Sexual partners within the past 3 months should be notified as the incubation period is up to 90 days.
- Partner notification may have to extend to 2 years for patients in secondary syphilis with clinical relapse in early latent syphilis.
- 40-60% of contactable sexual partners of patients with early syphilis also have the infection. Vertical transmission during pregnancy can occur at any time of pregnancy and with all stages of syphilis
- Epidemiological treatment for asymptomatic contacts of early syphilis should be considered unless partners are able to attend regularly for exclusion for syphilis.
- Serological tests for syphilis should be performed at the first visit and repeated at 6 weeks and 3 months.

III. Acquired Immunodeficiency Syndrome

Those who are diagnosed to be HIV positive should be referred to the HIV focal point which will which will be responsible for the rest of the actions.

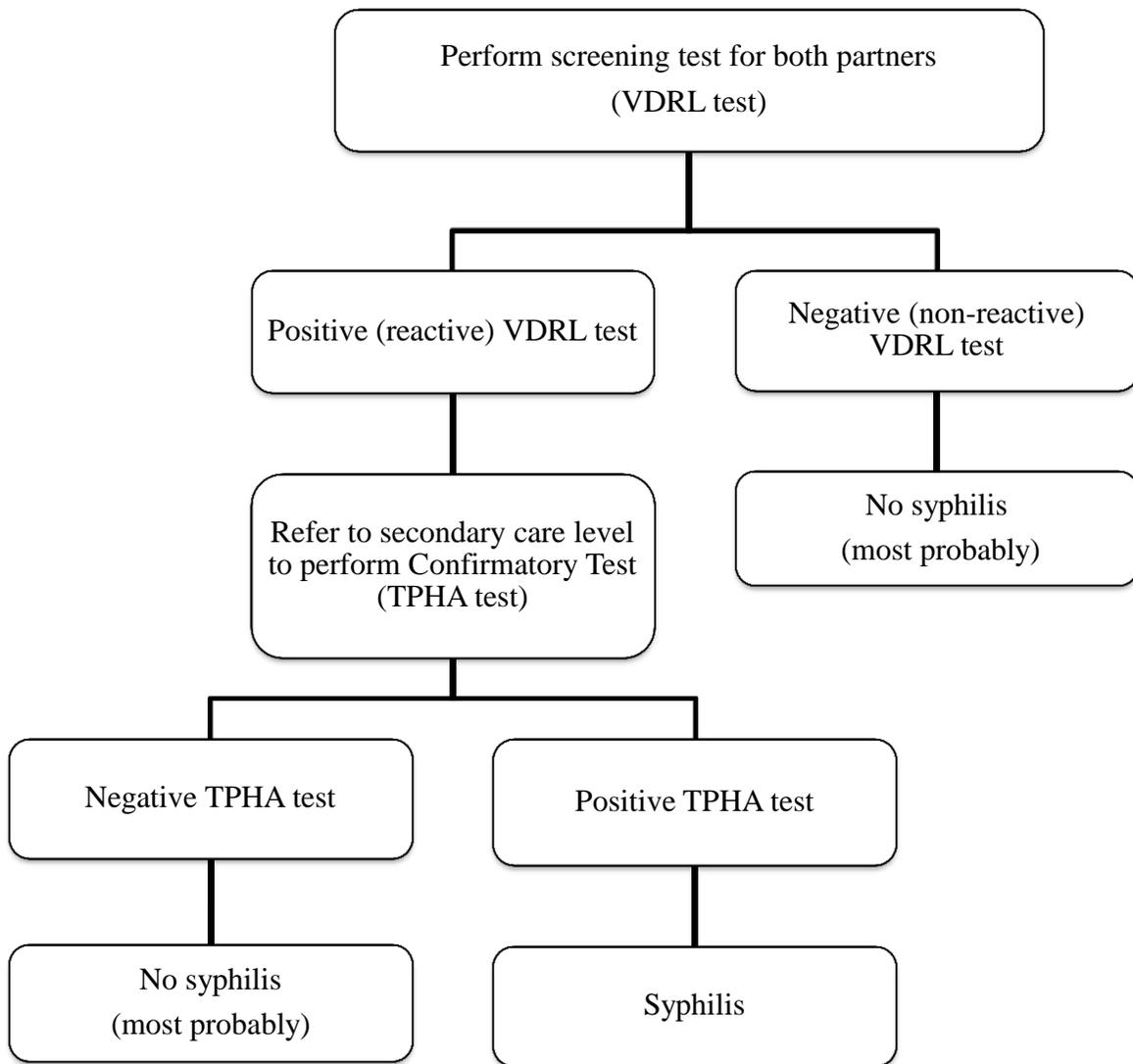


Figure 2: Premarital Counseling Protocol for Syphilis

Annex 1
Premarital Counseling Risk Assessment Form

جمهورية العراق
وزارة الصحة

رقم التسلسل

اسم المؤسسة الصحية
العدد:
التاريخ: 20 / / م

استمارة فحص المقبلين على الزواج

صورة الزوجة

صورة الزوج

الأسم:
العمر:
المهنة:
العنوان:
درجة القرابة بين الزوجين:

ثنائية ثالثة ابعد لاتوجد قرابة

التاريخ الصحي للعائلة:
أمراض الدم الوراثية:
التلاسيميا
فقر الدم المنجلي
الهيموفيليا
العوق الذهني
العوق البدني الولادي

الملاحظات

لا نعم لا نعم

الفحوصات المختبرية:
B-group & RH
VDRL
HIV
MCV
MCH
HBS Ag
TB

الاجراءات: تعطى الزوجة جرعة من لقاح توكسيد الكزاز ويؤشر الحقل الخاص به

توقيع الزوجة توقيع الزوج

ختم المؤسسة الصحية اسم وتوقيع مدير المؤسسة اسم وتوقيع الطبيب الفاحص

ملاحظة: لايجوز تسليد الاستمارة الا عند حضور الزوجين وبطرف مغلبي ومختوم بختم المؤسسة الصحية

Annex 2
Performance Checklist

I. Preparations for Premarital Counseling (PC)			
Task	Achieved		Comments
	Yes	No	
1. Trained physician on PC			
2. Trained paramedic on PC			
3. Specific place for PC			
4. Specific record for PC			
II. First Visit (to Paramedic/Health Worker)			
Task	Achieved		Comments
	Yes	No	
1. Welcome the couple			
2. Explain your plan of action			
3. Determine the degree of consanguinity between the couple			
4. Obtain the family history for both partners			
5. Draw blood sample from both partners for requested lab investigations			
6. Giving dose of tetanus toxoid to female partner			
7. Arrange next appointment for counseling			
8. Take phone numbers of couple			

III. Preparations between the Visits			
Task	Achieved		Comments
	Yes	No	
1. The paramedic reviews the family history of couple			
2. The paramedic reviews the results of laboratory investigations of couple			
3. The Paramedic discuss abnormal finding with the physician			
4. Rearrange separate appointment for each partner in case of abnormal findings			
IV. Second Visit (to the Physician)			
Task	Achieved		Comments
	Yes	No	
1. Welcome the couple			
2. Explain your plan of action			
3. Review the PC risk assessment form			
4. Take the medical history of each partner			
5. Perform the physical examination for each partner			
6. Assess the condition of each partner			
7. Make the required counseling			

Annex 3

Preconception Counseling

Preconception care and counseling should be provided to all women of childbearing age before and in-between pregnancies. Premarital visits represent a good opportunity for offering these services to improve maternal and neonatal outcome.

General Components of Preconception Counseling:

1. Health Education:

Health education should be given to all women concerning the need for antenatal care, the need to avoid the use of drugs (particularly in early pregnancy), and the importance of tracking her menstrual cycle.

2. Nutritional Counseling:

- Iron and Calcium: A particular need during pregnancy is to replenish the iron and calcium stores, either through good dietary habits or supplementation.
- Folic Acid: An adequate amount of folic acid is also essential. There is evidence that periconception intake of folic acid reduces the risk of congenital neural tube defects. No benefit is gained if the use starts after the first six weeks of pregnancy. A daily supplementation of 0.4 mg/day has been recommended for all women capable of becoming pregnant.
- Vitamin A: An excess intake of vitamin A from supplementary sources is to be avoided and stopped because of a possible link to a higher incidence of congenital anomalies.

3. Drugs and Special Habits:

- Preconception counseling offers an opportunity for health education of women to quit the use of certain drugs shown to be unsafe for conception.
- Smoking & Alcohol: Tobacco is now the leading preventable cause of low birth weight in many countries. Also, alcohol is well known to be a teratogen.
- Many women understand the risk of substance exposure during pregnancy, but they are generally unaware of the importance of avoiding exposure in the critical early weeks of pregnancy.

4. Testing for Rh and Blood Group:

Rh-factor and blood group typing should be done for all women awaiting the first pregnancy. If Rh negative, husband should also be tested. When the husband is also Rh negative, no risk is expected for incoming babies.

However, if the husband is Rh positive then special interventions should be undertaken to avoid Rh disease of the newborn. The woman should be informed of the need to have an injection of Rh immune globulin (Anti-D) during the second trimester; after abortion, amniocentesis, or abdominal injury during pregnancy; and within few days from birth of Rh-positive baby.

5. Rubella Vaccination:

Congenital rubella syndrome can be prevented by preconception screening and vaccination. Vaccination should be done at least three months before pregnancy. A woman with no history of vaccination and no contraindications to vaccination can be vaccinated without prior serologic testing.

6. Birth Planning:

- With the availability of modern effective methods for family planning, women can plan their birth to the optimal time from a health and social point of view. Births that are too early, too close, too late, and too numerous are better avoided.
- While an adolescent girl will be advised to postpone the first pregnancy, a woman over the age of 35 will be advised against postponing a pregnancy, since her chances of conception and successful outcome will, decrease with age.

Annex 4

Appropriate Age for Marriage of Women

Most studies have revealed that conception before the age of 18 years carries risks for both the mother and fetus. The marriage means subjecting the woman to the probability of being pregnant. This requires full maturation of the skeleton and reproductive system in order to protect the wife and her expected baby from many health problems, e.g. obstructed labor, premature delivery, spontaneous abortion, or pregnancy toxemia. The bones of pelvis in the young woman are not completely ossified which make them more prone to osteomalacia and deformities with increase in the likelihood of dystocia in the subsequent pregnancies.

In the other side, late marriage and conception beyond the age of forty can increase the probability of having children with chromosomal abnormalities, e.g. trisomy 21 and trisomy 13, 18, X and Y. The risk of the birth of child with Down syndrome is 1:1250 at age of 25 years, while it is 1 in 50 at age of 43 years. Additionally, the risk of all trisomes increases from 1:476 at age 25 years to 1:33 at 43 years.

Annex 5

Consanguineous Marriages (Endogamy)

Consanguineous marriages increase the probability of having children affected with hereditary diseases. The child carries the genetic traits of his/her parents whether desirable or not, e.g. vulnerability for certain hereditary diseases. The probability for having such diseases increases as the degree of consanguinity increases between parents, e.g. between sons of uncles, and/or aunts. The laws of probability indicate that every pregnancy may end with the birth of unhealthy infant. The figure is 2% in the case of marriage between foreigners and elevated to 4% in consanguineous marriages, i.e. doubled. Thus, it is important to take the family history for both partners, especially in the case of consanguineous marriage, and giving the appropriate counseling accordingly.

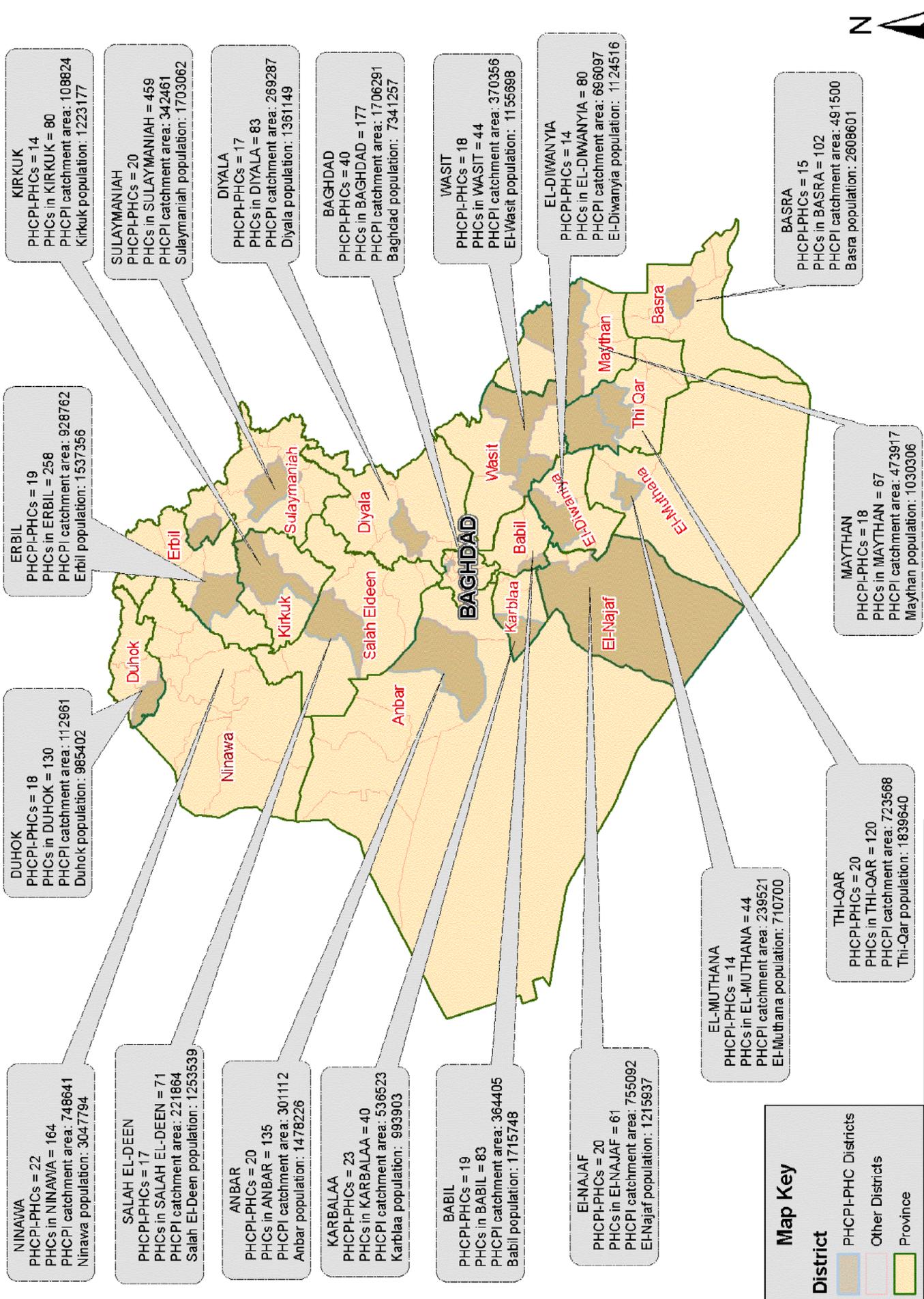
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PHCPI-PHCs population mapped to IRAQ population



Map Key

District

- PHCPI-PHC Districts
- Other Districts
- Province

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