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"The Effect of Sickle Cell Anemia on the Immune System in Najran Region Patients"

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Abstract:

Sickle cell anemia is an inherited disorder that affects red blood cells, especially hemoglobin, which lead to loses its function in carrying oxygen throughout the body, and change RBCs shape from normal (biconcave discs) to sickle or crescent shape. According to what was published by the Saudi Ministry of Health on its website in 2019, there are about 4.2% of the Kingdom's population with sickle-cell trait, and about 0.26% of them suffer from the disease (7). It was found that the pathophysiology of sickle cell disease includes many factors other than red blood cells, include immune cells (10). Because the patients they have a sickle cells which causes chronic vaso-occlusion this leads to dysfunction of spleen and during infection it's becomes to defect immune cells production which This leads poor protection against infection. In this study we studied the effect of sickle cell anemia on the immune system in patients in Najran region, and we found that it affects some immune cells through cell counts only, but we cannot prove this through the counts only, but rather its activity must be known.

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Abbreviation	the meaning
SCD	Sickle cell disease
SCT	Sickle cell trait
HbSS	Sickle cell haemoglobin
TAMs	tumor-associated macrophages
CBC	Complete blood count
RBCs	Red blood cell
WBCs	White blood cell
Hb	Haemoglobin
APC	Antigen presenting cell

NK	Natural Killer (NK) cells
P 1,2,3…ets	Patients

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1. Introduction :

Sickle cell anemia is an inherited disorder that affects red blood cells, especially hemoglobin, which lead to loses its function in carrying oxygen throughout the body, and change RBCs shape from normal (biconcave discs) to sickle or crescent shape. which leads to hemolysis and clots inside the vessels. It occurs more commonly in children (6,4). It effects of black African and Afro-Caribbean descent, and also those from the Mediterranean, Middle East, and parts of India (5). According to what was published by the Saudi Ministry of Health on

its website in 2019, there are about 4.2% of the Kingdom's population with sickle-cell trait, and about 0.26% of them suffer from the disease (7).

1.1 Sickle cell trait

Sickle cell trait is the heterozygous (having two different genes) form of sickle cell anemia, whereby the indi-vidual has both Hb-S and Hb-A in the RBCs. This condition is benign, as it is not a disease or anemia and the appearance of red blood cells is normal when seen in the blood film (8).

1.2 Sickle cell hemoglobin (HbSS)

The main function of haemoglobin is to carry Oxygen to the tissues and to return carbon dioxide (CO2) from the tissues to the lungs. the structure of normal haemoglobin (HbA) consists of four polypeptide chains is two α-globin chains and two β-globin chains , but there are disorders of hemoglobin and the most common of them are Sickle cell haemoglobin (HbSS) in the Sickle cell anemia patients, It happens when replacing thymine by adenine on 6th codon for the beta globin gene, which lead to change glutamic acid production to valine **(fig.1.2)** (6). Which then forms hydrophobic interactions with adjacent chains. Then resulting polymers align into bundles, causing change of the RBC shape to a crescent or sickle shape and reducing flexibility of passage of the cells into the blood vessels (5).

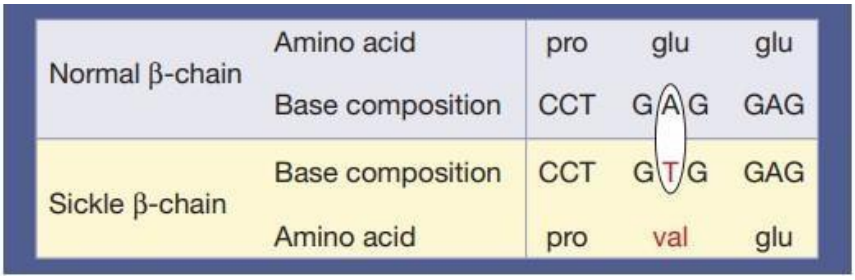

Figure 1.2 Molecular pathology of sickle cell anemia (8)

1.3 Immune system

The immune system is a complex collection of cells, tissues, organs, and proteins released by cells and that help the body fight infections and other diseases. This group contains white blood cells, organs and tissues of the lymph system, such as the lymph nodes, bone marrow, lymph vessels, thymus and spleen thymus (11). The immune system also plays an important role in eliminating tumor antigens and cancer cells. The immune response can be divided into two types, specific response to a specific antigen and a non-specific response. An important feature of a particular immune response is that it responds quickly to the antigen during the second exposure to the specific antigen. It is the memory of the initial response, and it is can also be divided into humoral responses and cellular response. humoral responses are antibodies produced in response to a given antigen, and these antibodies are proteins, have similar structures, and can be divided into various classes of immunoglobulins. Cellular responses are established by cells and can only be transferred by cells (11,17)

1.3.1 Innate immune system

the immune system consists of two types , the first one called innate immune system is the first line of defense against external threats through an inherent ability to recognize and rapidly respond to a broad range of pathogens and other immunogens, and by promoting the process of inflammation. Consists of many cell types and soluble molecules in tissues. The innate immune response incorporates three processes: 1- inflammation, produced by granulocytes and cytokines; 2- antibody response, produced by complement proteins; and 3- cellular response, facilitated by leukocytes and natural killer cells (11).

1.3.2 Adaptive immune system

the second one from immune system types called adaptive immune system. In contrast to the innate immune system, the more specific adaptive (acquired) immune system must be triggered by a specific virus, bacterium, or other foreign material, which stimulates lymphocytes to produce antibodies that can combat the foreign substance (18). At the next exposure, adaptive immunity is initiated by antigen-presenting cells (APCs), such as macrophages or dendritic cells, which help the immune system differentiate between the host's own cells (self) and those of invading bacteria or viruses. (11)

1.3.3 Immune cells

Monocyte

it is one of a type of immune cell which is generated in the bone marrow and moves through the blood to tissues , where it called macrophage or a dendritic cell when it in the tissues. Macrophages ingest microorganisms or foreign material and reinforce immune responses (8). The monocyte presenting on its surface the antigens to other immune cells during inflammation. A monocyte a type of WBCs which called phagocyte. During tumors monocytes may become tumor-associated macrophages (TAMs) at site of tumor. Monocytes are considered to be the largest white blood cells which measuring between 16 and 22 µm in diameter. Have one large kidney shaped nucleus (11).

- **Neutrophils**

Neutrophils are anther one of type of immune cells that helps treatment damaged tissues and fights infections (11). And it is one of the polymorphonuclear cell family. Increase levels of the neutrophil blood naturally its be in response to infections , injuries, or other of stress (17). This is done by releasing enzymes that kill microorganisms low number of neutrophils may be indicative of leukemia, some infections and vitamin B12 deficiency. It also regulates the immune response, and neutrophils have a lifespan of approximately 8 hours in the bloodstream (8).

- **Eosinophils**

Eosinophils have a nucleus with tow lobes and granules cytoplasmic. have a little number in the blood but may produce more of them in response to fight of parasitic and fungal infections or allergic disorders so its play important

role of immune response. also eosinophils are may be more large numerous in SCD patients than in healthy individuals and appear an activated state. However, it may be harmful in the inflammatory process of allergic diseases (8,11).

- **Basophils**

Basophils are anther one of a type of immune cell that is called granulocyte. its contain granules, and the granules contain histamine, heparin, and other molecules that secreted to fighting of infectious germs. Become to mast cells when it migrate to the tissues. Basophils help in prevent blood clot because it contain heparin. Also, an increase in the number of basophils can lead to itching as a reaction to allergies and thus may lead to complications such as enlarged spleen, bleeding , infections or cancer (11) .

- **Lymphocytes**

Lymphocytes are main type of immune cells that play important role in immune response of viruses. And it is exist in the blood flow and lymph tissue. There are divided to three types of lymphocytes is B-cell , T-cell and Natural Killer (NK) cells (8).

- **T- cell**

T lymphocytes is one of important type of cellular immunity. It is found in the periarteriolar sheath of the spleen and They are found in larger paracortical areas of lymph nodes. It also represents constitute 60-75% of type of lymphocytes . About 60% of mature T cells is CD4 called Helper and 30% is CD8 called Cytotoxic. It was also found to be normal in cases of sickle cell disease (4). T cells have an important role in the specific immune response to antigen (11).

- **B-cell**

if the B- cell remain for long time in the body it's called memory cells. This causes them to remembers the receptors on the surface of previous antigens, helping the immune system to respond more faster. Lymphocyte B cells can be found in primary lymphoid tissue such as bone marrow and secondary lymphoid tissue such as spleen and lymph nodes. The B-lymphocyte response to the original antigen is done by the BCR. It can also differentiate into plasma cells that produce antibodies after activating mature cells by binding of BCR with a specific antigen (11,8).

- **Natural Killer (NK) cells**

Natural Killer cells are one of importunes immune cell that considered from lymphocytes family. The virally infected cells are killing by NK , and early signs of cancer are detecting and controlling by NK. NK cells express various types of immunoreceptors that are all designed to sense pathological changes of self cells. And it was found that there is an absolute increase of NK in SCD patients not receiving hydroxyurea therapy (16).

Adults	Blood count	Children	Blood count
Total leucocytes	$4.00–11.0\times10^9$/L*	*Total leucocytes*	
Neutrophils	$1.8–7.5\times10^9$/L*	Neonates	$10.0–25.0\times10^9$/L
Eosinophils	$0.04–0.4\times10^9$/L	1 year	$6.0–18.0\times10^9$/L
Monocytes	$0.2–0.8\times10^9$/L	4–7 years	$6.0–15.0\times10^9$/L
Basophils	$0.01–0.1\times10^9$/L	8–12 years	$4.5–13.5\times10^9$/L
Lymphocytes	$1.5–3.5\times10^9$/L		

* Normal black and Middle Eastern subjects may have lower counts. In normal pregnancy the upper limits are: total leucocytes 14.5×10^9/L, neutrophils 11×10^9/L.

Figure 1.3.3: white blood cells normal counts (8).

- **Mast cell**

The mast cell is type of immune cell and it is a large mononuclear cell it is have a small nucleus and contains heparin and histamine in their granules. Mast cell is an important in injury , infection and cellular immunity generally. It can be found in tissues and around blood vessels. And mast cells are originally B cells that became transported into tissues and became active due to exposure of the body to the antigen. And It has been reported that mast cells are more active in sickle cell patients (14).

- **Macrophages**

Macrophages are one another type of immune cell and they are large mature monocytes found in the tissues (17). When the body is exposed to an infection, monocytes leave the bloodstream and into the tissues to become macrophages, where they can engulf the microorganism or infected cells. And thus the microbe's antigen is displayed on its surface to alert other immune cells to this antigen, and this process is called the acquired immune response(11).

- **Dendritic cells**

Dendritic cells are monocytes found in tissues and known as antigen-presenting cell (APC) (11). Dendritic cells play role of the initiation of adaptive immune responses. They are not only important for inducing primary immunity but may also be important for inducing immune tolerance (19).

1.4 The effects sickle cell disease on immune system

It was found that the pathophysiology of sickle cell disease includes many factors other than red blood cells, such as immune cells (e.g. macrophages , mast cells monocytes, neutrophils, eosinophils, basophils and natural killer cells) (10).

Because patients with sickle cell disease have the abnormal shape of red cells (sickle cells). This causes chronic vaso-occlusion which leads to dysfunction of spleen and during infection it may result in defect in adaptive immune cells production. This leads poor protection against infection. Another defect was found is in complement activation, in which the cascade can be activated either via the classical pathway, following binding of IgM or IgG to surface antigens, or the alternative pathway, in which C3b interacts directly with the pathogen cell surface, then recruiting further downstream components. Some studies and experiments were conducted in vitro to verify the activity of the complement components, and it was found that there is a functional deficiency in the activity of the alternative pathway with lower levels of the active form of factor B

(the first protein recruited by C3b) and impaired opsonization (5). Also, patients with SCD also have cell-mediated immune disorder, which is related to zinc deficiency. The low level of zinc in sickle cell patients may be a contributing factor in susceptibility to infection. that leads to reduce T-cell production (5,20). On other hand, it was reported that any person has Sickle Cell Trait (SCT) showed to be protective against to Plasmodium falciparum malaria infections. The HbS often associated with a decrease parasitic invasion to RBCs that lead to weak multiplication, and accelerated remove of malaria by the spleen. Also because of the parasite invading red blood cells will produce intracellular hypoxia that lead filtration of parasitized cell by spleen (5).

1.4.1 Monocyte

In one study it was found that monocytosis in sickle cell patients is common and correlates positively with hemolysis and negatively with hemoglobin. It was also found to be lower in children with sickle cell disease who were treated with hydroxyurea (10).

1.4.2 Neutrophils

It was also found that neutrophils is high and involved in the pathophysiology of sickle cell disease patients and positively correlated with sickle cell anemia (10).

1.4.3 Eosinophils

Eosinophils have also been reported to be higher in patients with sickle cell anemia compared to healthy controls, and it was also more active even in the absence of parasitic infections (10).

1.4.4 Basophils

In one study that consisted of 54 patients with sickle cell disease and 27 healthy controls, it was found that basophils was normal in both groups, indicating that there was no effect on them (12,10).

1.4.5 Lymphocytes

One study that examined the association of CD4 T-cell in sickle cell anemia showed no significant difference of CD4 T-cell in the sickle cell anemia patients in comparison to healthy individuals (4). In another study that was talking about " B-cell changes occur in patients with sickle cell anemia " was shown that there were no significant changes in the percentage of B-cells. However, the results of in vitro experiments showed that changes in an that B-cells function occur during vaso occlusive pain crises in patients with sickle cell anemia (13). Moreover, regarding NK cells, it has been found that the cytotoxicity of NK cells is significantly increased compared with healthy individuals (10). Also a study conducted on the effect of hydroxyurea treatment in sickle cell patients on NK cells found that there is an absolute increase of NK cells in SCD patients who were not receiving hydroxyurea therapy (16).

1.4.6 Mast cell

In a study conducted on mice, it was found that mice with sickle cell pathobiology have more mast cells activation than healthy mice (14). It was also found in another study that histamine correlated negatively with hemoglobin F and positively with the activity of mast cells. This indicates mast cell activity rather than an increased number of mast cells in sickle cell patients, indicating the functional activity of mast cells (10).

1.4.7 Macrophages

In many studies (9,10,15) conducted on mice which found that macrophages may cause inflammation leading liver damage, hepatocyte apoptosis and fibrosis in the mice with sickle cell anemia (15). This may be because of IL-1β production in SCD due to that heme induces IL-1β processing through NLRP3 inflammasome activation in macrophages (9,10)

1.4.8 Dendritic cells

Dendritic cells play a role of the initiation of adaptive immune responses and in causing orthopedic complications associated with sickle cell anemia in sickle-cell patients (17). It has also been found that there is gene overexpression of the bone morphogenetic protein-6 of dendritic cells derived from monocytes in sickle cell patients (10).

2. Objective

- A study of the causes of effects sickle cell anemia on immune system in patients at Najran region.

3. Materials:

- We collected data for sickle cell anemia patients from (King Khalid Hospital and Najran General Hospital
- We Used Microsoft excel 2010 program for data analysis .

4. method :

4.1 we collected 10 random data for sickle cell anemia patient include : 6 males and 4 females from (King Khalid Hospital and Najran General Hospital).

- After that, we entered the complete blood counts (CBC) results (WBCs, hemoglobin) and differential white blood cells count (monocyte , neutrophils , eosinophils , basophils and lymphocytes) into the Microsoft excel 2010 program.
- After that we did a statistical analysis by using columns graph tool.

5. Results:

The mean age of the patients was **± 8** years and their distribution according to sex was as follows:

Patients number	Sex
P1	Female
P2	Male
P3	Male
P4	Female
P5	Male
P6	Male
P7	Male
P8	Male
P9	Female
P10	Female

Table 5.1: sex of the patients

the normal range of the tests was as follows :

Test	Normal range
Hb	M: 12 – 17 … F: 12 – 16 g/dL
WBCs count	9.1 – 28.7 x10^3/uL
Monocyte	0.3 – 1.5 x10^3/uL
Neutrophils	1.9 – 11.5 x10^3/uL
Eosinophils	0.05 – 0.5 x10^3/uL
Basophils	0 – 0.2 x10^3/uL
Lymphocytes	6.7 – 14 x10^3/uL

Table 5.2: the normal range of the tests. Adopted from Najran Regional Lab.

5.1 Hemoglobin results:

There was significant decrease of hemoglobin in both females and males. All Patients had (HbSS) and decreased (HbA) Figure 5.1 .

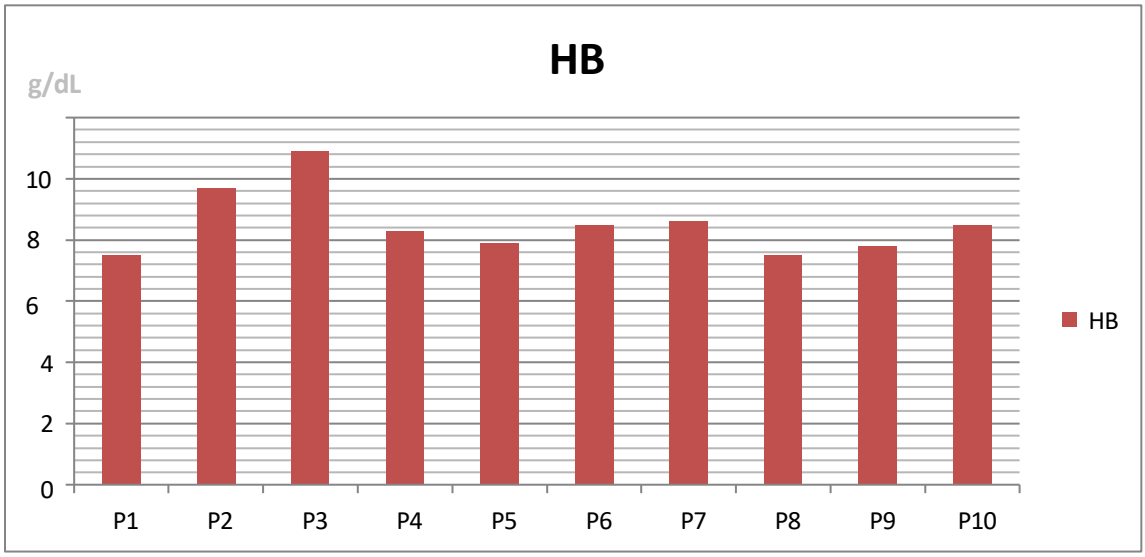

Figure 5.1 Results of hemoglobin (P1 ,P4,P9,P10)is females and (P2,P3,P5,P6,P7,P8) is males Normal range (M: 12 – 17 … F: 12 – 16 g/dL)

5.2 WBCs count :

All the results of WBCs count were within the normal range except P3 was low Figure 5.2.

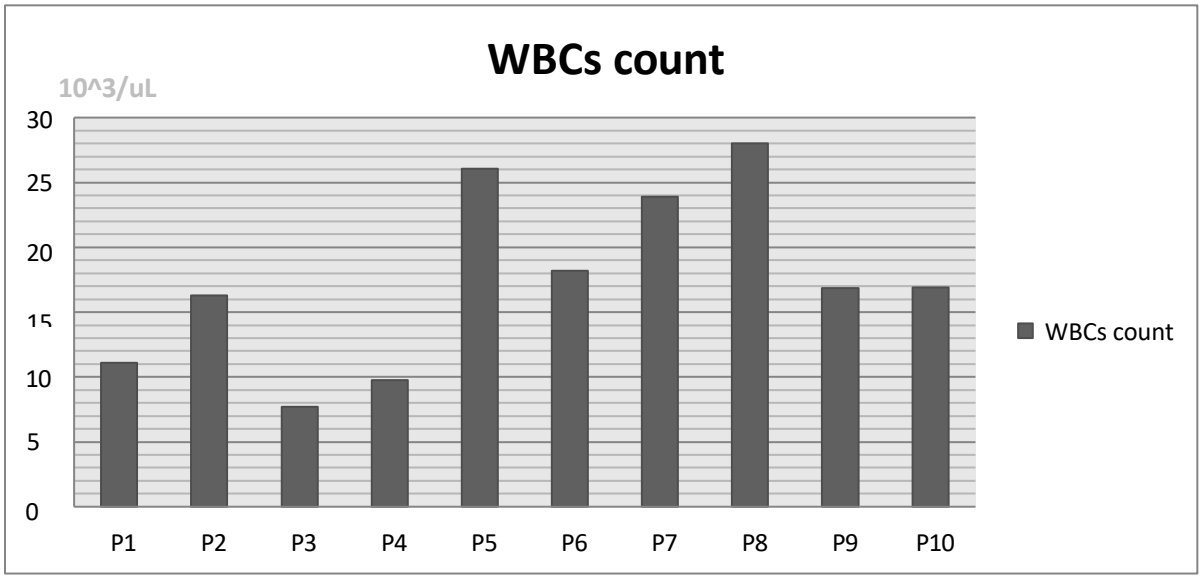

Figure 5.2: results of WBCs count (P1 ,P4,P9,P10)is females and (P2,P3,P5,P6,P7,P8) is males. Normal range (9.1 – 28.7 x10^3/uL)

5.2.1 Monocytes count :

There was significant increase of monocyte in males , but the females

were within the normal range **Figure 5.2.1 .**

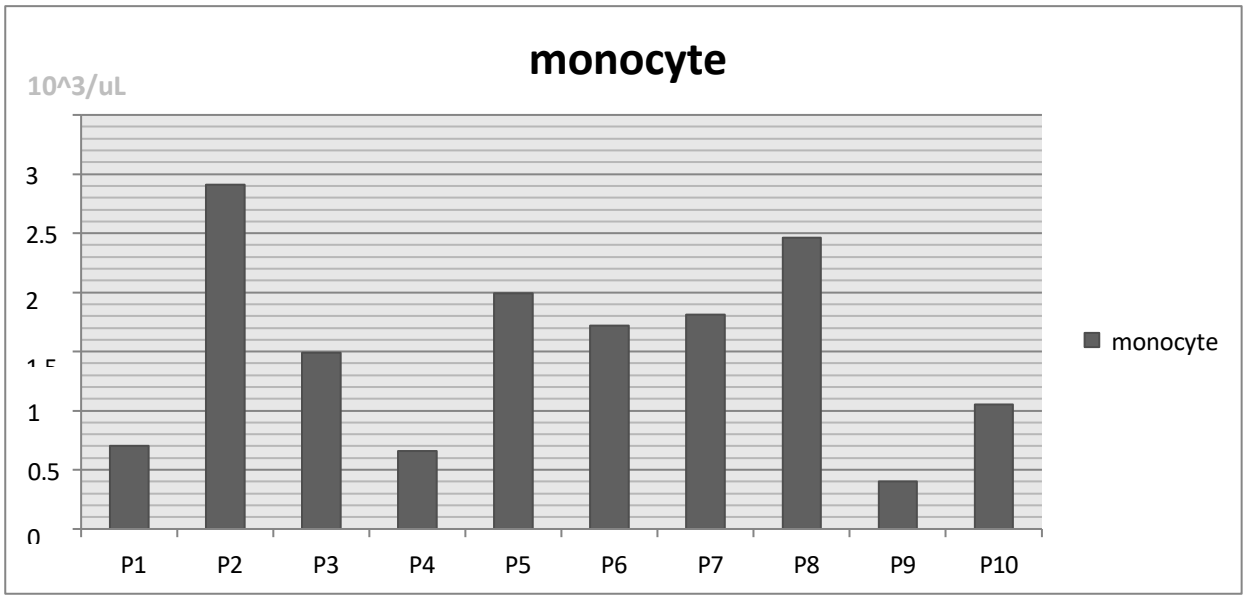

Figure 5.2.1 : results of monocytes count (P1 ,P4,P9,P10)is females and (P2,P3,P5,P6,P7,P8) is males. Normal range (0.3 – 1.5 x10^3/uL)

5.2.2 Neutrophils count :

There was significant increase of neutrophils count in P6, P7 and P9 , but others patients were within the normal range Figure 5.2.2 .

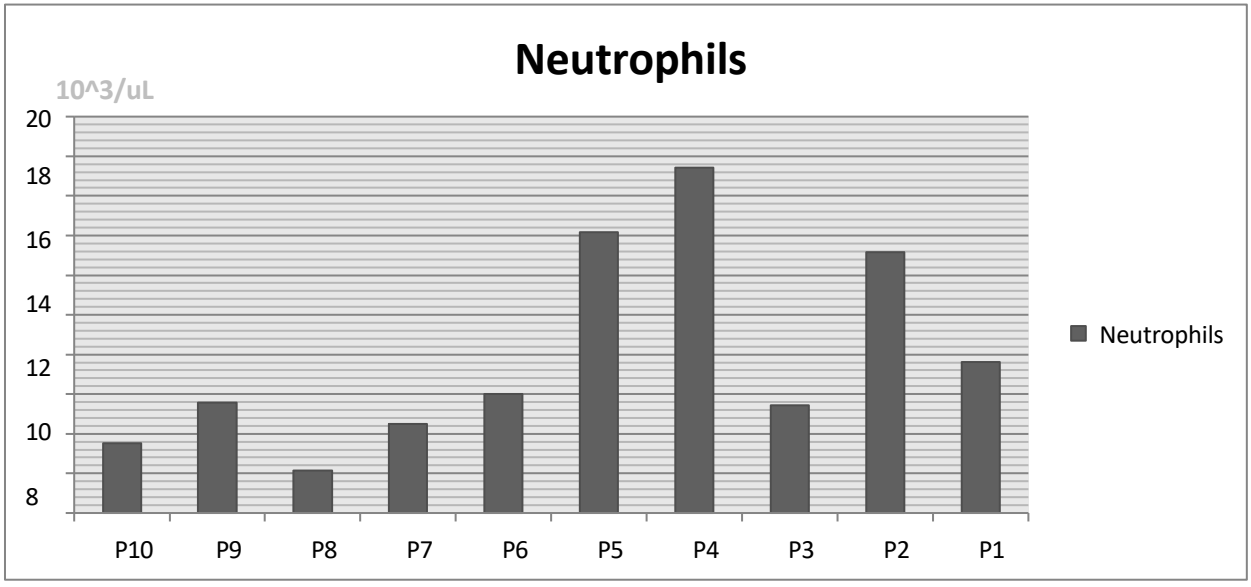

Figure 5.2.2: results of Neutrophils count (P1 ,P4,P9,P10)is females and (P2,P3,P5,P6,P7,P8) is males. Normal range (1.9 – 11.5 x10^3/uL)

5.2.3 Eosinophils count :

There was significant increase of eosinophils count in P4 and P5 , but other Patients were within the normal range Figure 5.2.3 .

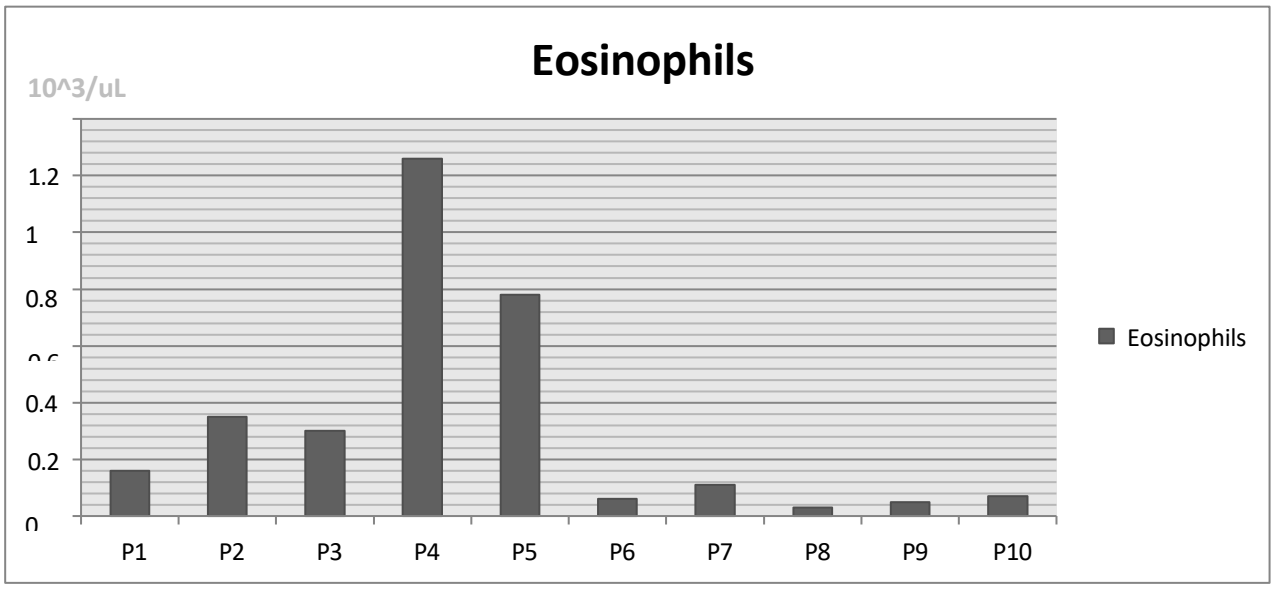

Figure 5.2.3: results of eosinophils count (P1 ,P4,P9,P10)is females and (P2,P3,P5,P6,P7,P8) is males. Normal range (0.05 – 0.5 x10^3/uL)

5.2.4 Basophils count :

There was significant increase of basophils count in P5 and P7 , but other Patients were within the normal range Figure 5.2.4 .

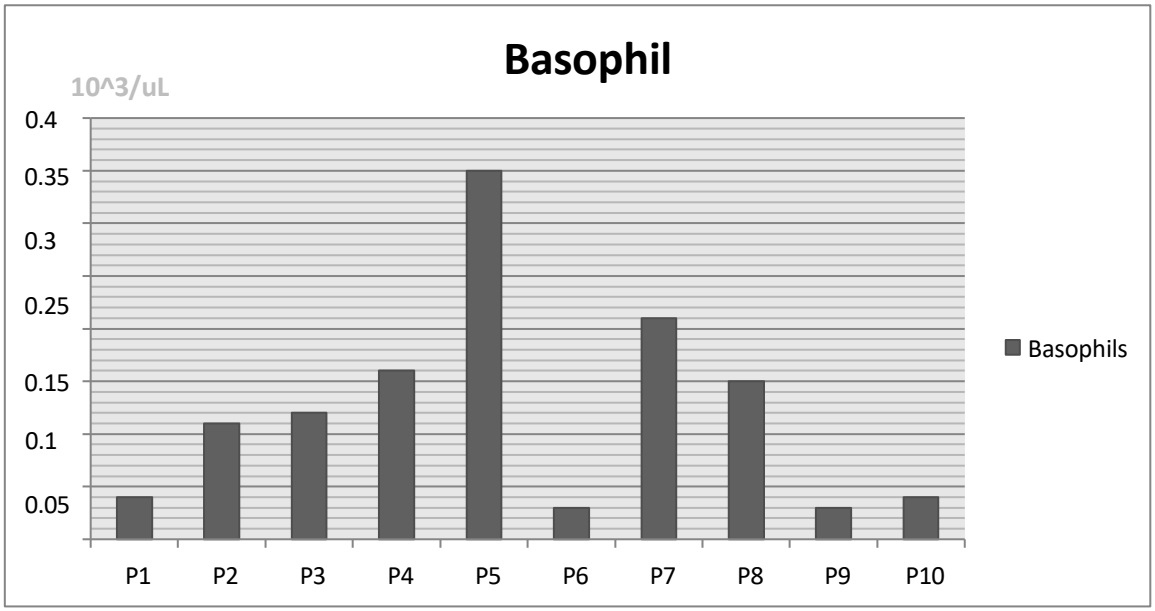

Figure 5.2.4: results of basophils count (P1 ,P4,P9,P10)is females and (P2,P3,P5,P6,P7,P8) is males. Normal range (0 – 0.2 x10^3/uL)

5.2.5 lymphocytes count :

There was significant increase of lymphocytes count in **P5** and **P8** and significant decrease in **P3** , **P4** , **P6** , **P7**, and **P9** and within the normal range in **P1** ,**P2** and **P10 Figure 5.2.5 .**

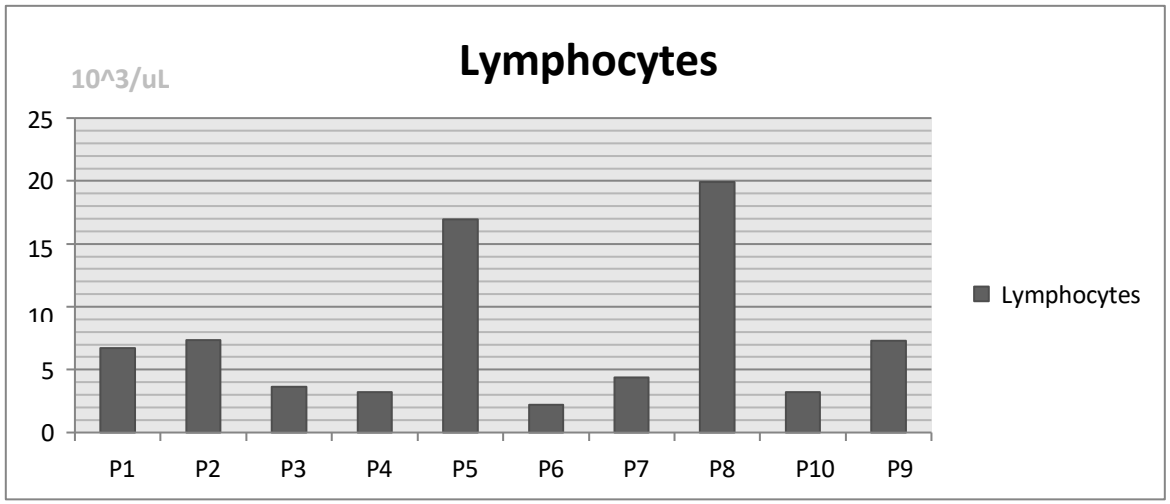

Figure 5.2.5: results of lymphocytes count (P1 ,P4,P9,P10)is females and (P2,P3,P5,P6,P7,P8) is males. Normal range (6.7 – 14 x10^3/uL)

5.3 Average of normal range:

All the mean of the results of tests were within normal range. As can be seen in **Table 5.3**, statistical analysis of the mean showed no Signiant difference between all tests, indicating that most the effects is on the physiological level.

Test	Mean	Normal range
Hb	M : ± 8.9 g/dL … F : ± 8.0 g/dL	M: 12 – 17 … F: 12 – 16 g/dL
WBCs count	± 17.47 x10^3/uL	9.1 – 28.7 x10^3/uL
Monocyte	± 1.5 x10^3/uL	0.3 – 1.5 x10^3/uL
Neutrophils	± 7.95 x10^3/uL	1.9 – 11.5 x10^3/uL
Eosinophils	± 0.32 x10^3/uL	0.05 – 0.5 x10^3/uL
Basophils	± 0.12 x10^3/uL	0 – 0.2 x10^3/uL
Lymphocytes	± 7.48 x10^3/uL	6.7 – 14 x10^3/uL

6. Discussion:

In our analysis, we found that there are different effects of sickle cell disease on immune cells. Compared to previous researches, we find that there is agreement in some points between our analysis and previous studies. On the other hand, we find that there is a big difference between them.

In our analysis, we find a significant decrease in hemoglobin with an increase in (HbSS) and decreased (HbA). These results are supported by previous findings that proved change of glutamic acid production to valine (8). Thus, patients with sickle cell have hemoglobin SS.

We also find increased neutrophils is common in some patients. These results are supported by previous findings that proved also the increase of neutrophils which positively correlated with sickle cell disease severity (10).

monocytes were also found to be in our analysis male showed monocytosis but for females, they were within the normal range. Previous studies reported increased monocytes in patients with hemolysis regardless the gender (10). This explains to us a possible explanation that the monocytes are not associated with hemolysis or hemoglobin, but rather are affected by gender, as the cause has not been identified. Therefore, more analyzes may be needed to find the relationship and exact description of their association with sex.

The eosinophils in the results shown to us, we found that they were not affected in most of the sickle cell patients. On the other hand, previous studies do not support our results, that showed the eosinophils to be high in sickle cell patients (10)

Basophils shown in the results of our analysis were normal in most patients. These results are supported by previous research that showed normal basophils, and this proves that basophils are not affected by sickle cell disease (12).

In our analysis, we found that the count lymphocyte varied between high, low and within the normal range, and this explains to us that the of effect of SCD on lymphocytes cannot be determined by just count ,but rather activity must be known in order to determine the effect of SCD on lymphocytes. These results are supported by previous findings that showed a variation in the count of lymphocytes (4,10).

In our analysis, we found that there are variations in the numbers of immune cells for all patients and that they may suffer from infections of all kinds, and this may pose a greater risk to their health. Also these results are supported by previous results that SCD patients are susceptible to infections (1,5,10).

7. Conclusion:

In our study, we found that sickle cell disease affects some immune cells such as monocytes, neutrophils, and lymphocytes, but we cannot prove that effects through cell counts only, but rather their activity must be known as well. The counts of other immune cells such as NK, B-cell, T-cell and macrophages should have been included but due to limitations of data we could not. These are very important factors to have a clear idea about the effects of SCD on immune system. In addition, the number of data obtained for sickle cell patients was few.

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"تأثير فقر الدم المنجلي على الجهاز المناعي لدى مرضى منطقة نجران"

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الملخص:

فقر الدم المنجلي هو اضطراب وراثي يؤثر على خلايا الدم الحمراء، وخاصة الهيموجلوبين، مما يؤدي إلى فقدان وظيفتها في حمل الأكسجين في جميع أنحاء الجسم، وتغير شكل كرات الدم الحمراء من الطبيعي (أقراص ثنائية التقعر) إلى الشكل المنجلي أو الهلالي. بحسب ما نشرته وزارة الصحة السعودية على موقعها الإلكتروني

في عام 2019م، هناك حوالي 4.2% من سكان المملكة مصابون بداء فقر الدم المنجلي، وحوالي 0.26% منهم يعانون من المرض (7). وقد وجد أن الفيزيولوجيا المرضية لمرض فقر الدم المنجلي تشمل عوامل عديدة أخرى غير خلايا الدم الحمراء، وتشمل الخلايا المناعية (10). لأن المرضى لديهم خلايا منجلية تسبب انسداد الأوعية الدموية المزمن وهذا يؤدي إلى خلل في الطحال وأثناء الإصابة يصبح خلل في إنتاج الخلايا المناعية مما يؤدي إلى ضعف الحماية ضد العدوى. قمنا في هذه الدراسة بدراسة تأثير فقر الدم المنجلي على الجهاز المناعي لدى مرضى منطقة نجران، فوجدنا أنه يؤثر على بعض الخلايا المناعية من خلال تعداد الخلايا فقط، ولكن لا يمكننا إثبات ذلك من خلال التعدادات فقط، بل يجب نشاطه كن معروف.