

Sudan University of Science and Technology

College of Graduate Studies

**Study of Patients Abdomen Sicklier by Using
Ultrasound**

دراسة بطن مرضي الانيميا المنجلية باستخدام الموجات فوق الصوتية

**A thesis Submitted for Partial fulfillment of the Requirement of M.Sc
Degree in Diagnostic Medical Ultrasound**

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Date 2014

الاية

أَمَّنْ هُوَ قَانَتْ آنَاءُ اللَّيْلِ سَاجِدًا وَقَائِمًا يَحْتَزُّ
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أُولُوا الْأَلْبَابِ

الزمر آية ٩

Dedication

For my father who grants me power and support.

For my mother who gives me an endless hope and utmost love.

For my sweet heart and the light of my life.
My daughters Yumna and Sajedah .

Acknowledgement

I would like to thank God firstly for enabling me to complete this thesis and I want to express my sincere thanks to Dr. Elsafi Ahmed Abdulla the supervisor of my thesis .I am very grateful to ,all my teachers in all educational levels ,especially thanks for my teachers Dr. Caroline Edward, Dr. Ahmed Mostafa AbuKonna .

My great thanks to my colleges in Mohammed El Amin Hamid hospital for their help and support.

Special thanks to my parent for his patient and supporting me.

Finally all my thanks and grateful to my small family my husband and my kids .

Thanks to all

Abstract

This is a prospective study deals with study of patient's abdomen with sickle cell anemia Using ultrasound to detect the complication on the abdominal organs and to identify the correlation between the duration of the disease and its complications.

The area of the study was in Mohammed ELAmeen Hamid hospital in Omdurman. In this study 35 patient were investigated as a sample volume and 10 children were considered as a control group.

By the end of this study the researcher found that the SCD prevalence in Sudan specially western tribes between male and female, this hereditary disease affected whole the body like abdominal strong organs specially the spleen which is affected by the age (duration of disease) firstly become enlarged and after that start to shrink and in some cases disappear

(Autosplenectomy), about 31.4% of patient, 2.85% affected kidneys, 5.7% gall stones and

2.85% have jaundice. Lastly we found that the ultrasound exam is first choice to detect and follow the complication of SCD on abdominal organs.

الملخص

هذه دراسة توقعية تعني بدراسة بطن المرضى المصابين بمرض الانيميا المنجلية بواسطة استخدام الموجات فوق الصوتية ، لاكتشاف المضاعفات في الاعضاء البطنية ، لمطابقة الارتباط او العلاقة ، بين مدة المرض وهذه المضاعفات .

تمت هذه الدراسة بمستشفى محمد الامين حامد بامدرمان .

اجريت هذه الدراسة علي حوالي ٣٥ طفل مريض تم فحصهم ، وحوالي ١٠ اطفال تم اعتبارهم كمرجع .

في نهاية هذه الدراسة وجدت الباحثة ان مرض الانيميا المنجلية منتشر في السودان خاصة قبائل غرب

لسودان والمرض منتشر بين الذكور والاناث ، هذا المرض الوراثي يؤثر علي كل الجسم مثل الاعضاء البطنية الصلبة ، وخاصة الطحال ، والذي يتاثر بمدة المرض ففي بدايته يبدأ بالتضخم ثم بعد ذلك يبدأ بالانكماش في حجمه ، وفي بعض الحالات يختفي تماما ، وهنا كانت هذه الحالات حوالي ٣١.٤% اختفي طحالهم ، ٢.٨٥% ناثرت الكلي لديهم ، و ٢.٨٥% كانت لديهم اصابة باليرقان .

اخيرا وجد ان فحص الموجات فوق الصوتية هو الخيار الاول لاكتشاف مضاعفات مرض الانيميا المنجلية ومتابعتها .

List of abbreviation

SCA sickle cell anemia

SCD sickle cell disease

RBCs red blood cells

Pt patient

Hb hemoglobin

Table of contents

Content	Page
الإلية	i
Dedication	ii
Acknowledgment	iii
Abstract	iv
الملخص	v
List of abbreviation	vi
Table of content	vii
Table of content	viii

Chapter one

Introduction	1
Research problem	6
Objectives	6
Over view of the study	7

Chapter two

The literature review	8
Anatomy	8
Physiology	14
Pathology	16
Previous studies	21

Chapter three

Material and methods	24
Type of study	24
Population of the study	24
Study sample	24
Machine used	24
Technique	24
Data collection	25
Data analysis	25
Data storage	25

Chapter four

Results	26
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Chapter five

Discussion	37
Conclusion	39
Recommendation	40
References	41
Appendix	43

Chapter one

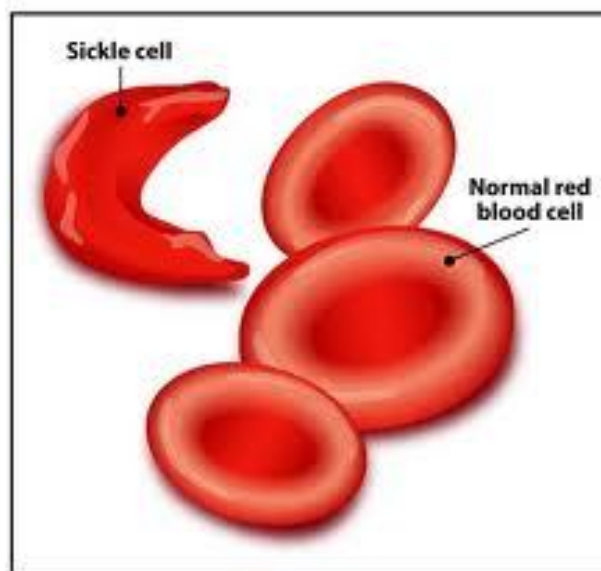
1.1 Introduction:

Sickle Cell Anemia (SS) is an inherited blood disorder (autosomal recessive). Approximately one in 400 black babies is born with Sickle Cell Anemia, and about one in Live has Sickle Cell Trait (AS).

The two hemoglobin types inherited will determine the shape of the red blood cell (RBC). When both parents have Sickle Cell Trait, there is a 1-in-4 chance (25 percent) the baby will have normal hemoglobin (AA), a 50 percent chance the baby will have Sickle Cell Trait (AS), and a 1-in-4 chance (25 percent) the baby will have Sickle Cell Anemia (SS). These chances remain the same with each pregnancy. (Debra A. Vedro, MSN, RN, CPNP And Rebecca A. Morrison, MSN, RN, CPNP)

Normal red blood cells are smooth, round and flexible, like the letter O, enabling them to move easily through your child's blood vessels. In contrast, sickle cells are stiff, sticky and often shaped like the letter C. Sickle cells tend to cluster together, making it difficult for them to move through small blood vessels. These clusters can create blockages in your child's small blood vessels, stopping the movement of healthy, oxygen-carrying blood. This blockage, which can be painful, is what causes the majority of the complications of the disease. (Matthew Heeney, MD, © Children's Hospital Boston, 2011)

Sickle cell anemia



Sickle cells only live for about 14 days, while normal red blood cells can live up to 120 days. This is because the spleen, which helps filter bacterial infections from the blood and act as the recycle center for old red blood cells, tends to destroy the sickle cells. This leads to a lack of oxygen-carrying red cells in the blood, causing chronic anemia. Sickle cells can also damage your child's spleen, increasing the risk of bacterial infection (Matthew Heeney, MD, © Children's Hospital Boston, 2011).

Sickle cell anemia is one of the major types of anemia found in Sudan, especially in western Sudan in which the sickle cell gene is frequent.

The SCA by its self not consider big problem but the complication of it grading from simple like (pain ,infections) to dangerous which lead to death.

In this study we want to show this complication of SCD which affected the abdominal organs by using ultrasound .

There was many study around the worlds deal with this matter (Ma'aji SM, et all 2012) ,(A.A Babadoko, 2012) ,(Al-Salem AH1,.1998) ,(Olatunji AA1, Olatunji PO. 2001) ,(Al-Salem AH1,.1998) , but in the Sudan this type of study is few like (Bakhieta Ibrahim Attalla, MD 2010) .

1.2 Types of sickle cell disease:

There are a number of different types of sickle cell disease that differ in symptoms and severity. The type of sickle cell disease depends on the specific genetic defect that your child has inherited.

1.3 Sickle cell trait (HbAS):

A person has an HbS gene and normal hemoglobin a gene (HbA). Under normal circumstances, these children have no symptoms of sickle cell disease.

1.4 Sick cell anemia (HbSS):

Child has two HbS genes; one inherited from each parent most common and most severe form of sickle cell disease variety of symptoms and complications of sickle cell disease occur severe, chronic anemia is common.

1.5 Sick cell with hemoglobin C disease (HbSC):

Child has one HbS gene and one defective hemoglobin C gene (HbC) mild to moderate anemia may occur. some complications of sickle cell disease may occur but to a milder degree.

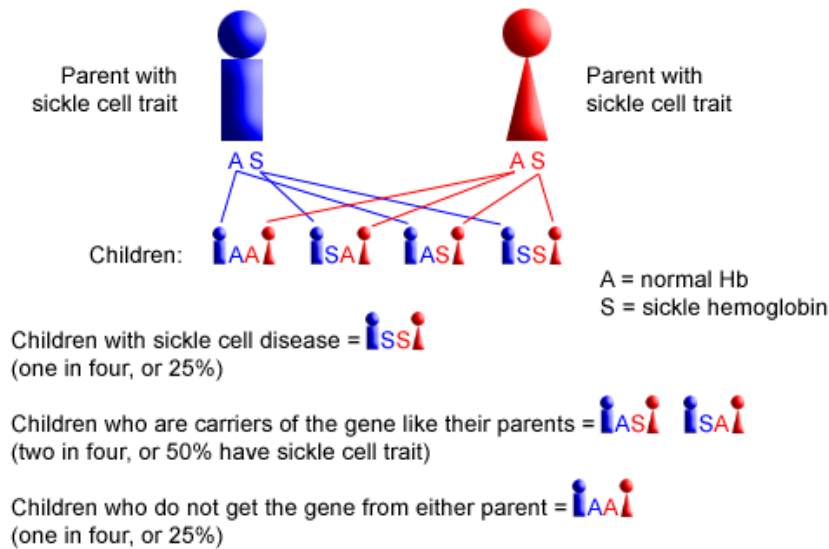
1.6 Hemoglobin S-beta-thalassemia:

Comes in two forms: sickle beta zero and sickle beta plus child has one HbS gene and one beta thalassemia gene mild to severe anemia may occur. Children may experience a broad range of sickle cell symptoms and disease severity.

Other forms of sickle cell disease (compound heterozygotes), including HbSE, HbSO and HbSD. child has one HbS gene and another abnormal hemoglobin gene symptoms vary depending on the specific genetic defect.

1.7 The most common ways a child can develop sickle cell disease:-

If both parents have sickle cell trait, each of their children will have 25 percent chance of having sickle cell disease. If one parent has sickle cell disease and the other has sickle cell trait, each of their children will have a 50 percent chance of having sickle cell disease and a 50 percent chance of having sickle cell trait. If one parent has sickle cell disease and the other does not, all of their children will have sickle cell trait, but none of them will have sickle cell disease.



It mostly affects children of African descent and Hispanics of Caribbean ancestry, but also is found in those with Middle Eastern, Indian, Latin American, Native American and Mediterranean heritage. Millions worldwide suffer complications from sickle cell disease; 2 million African-Americans, or 1 in 12, have the sickle cell trait. (Matthew Heeney, MD © Children's Hospital Boston, 2011)

1.8 Symptoms and complications:-

Each child may experience symptoms differently. The symptoms of sickle cell disease can vary greatly depending on the specific type of the disease.

1.8.1 Anemia

Most common symptom of all the sickle cell diseases if chronic, may delay healing, and normal growth and development Pain crisis (also called vaso-occlusive crisis) occurs when the flow of blood is blocked to an area because the sickle-shaped cells have become stuck in the blood vessel pain can occur anywhere but most often occurs in the bones of the arms,

legs, chest and spine painful swelling of the fingers and toes (dactylitis) can occur in infants and toddlers priapism, a painful sickling that occurs in the penis any interruption in blood flow to the body can result in pain, swelling and possible death of the surrounding tissue not receiving adequate blood and oxygen.

1.8.2 Acute chest syndrome

When sickle-shaped cells stick together and block the flow of oxygen in the tiny vessels in the lungs can be life-threatening often occurs suddenly, when the body is under stress from infection, fever or dehydration resembles pneumonia and includes fever and breathing symptoms such as cough or difficulty catching breath. Multiple episodes can cause permanent lung damage .Bacterial sepsis and infection most children, by toddlerhood, no longer have a functioning spleen. The spleen is important in the body's defense against serious bacterial infections; therefore, children with sickle cell are at risk for life-threatening bacterial infection. Fever is a symptom that must be evaluated immediately to rule out a life-threatening bacterial infection.

1.8.3 Splenic sequestration (pooling):

Pooling of sickle-shaped cells in the spleen, causing a sudden drop in hemoglobin can be life threatening if not treated promptly spleen can become enlarged and painful from the increase in blood volume spleen becomes scarred and permanently damaged after repeat episodes repeated episodes of sequestration requires surgical removal of the spleen

1.8.4 Stroke:

This is a sudden and severe complication that can occur in children with sickle cell disease.

Sickle-shaped cells can block the major blood vessels that supply the brain with oxygen.

Any interruption in the flow of blood and oxygen to the brain can result in devastating neurological impairment. Symptoms of a stroke can include: weakness, particularly on one side of the body; slurred speech, seizure, confusion, dizziness or loss of coordination, or severe headache having had one stroke, a child is much more likely to have more strokes and requires preventative therapy

1.8.5 Jaundice:

Common sign and symptom of sickle disease results from excess red blood cells being destroyed in the spleen. Any and all major organs are also affected by sickle cell disease. The liver, heart, kidneys, gallbladder, eyes, bones and joints can suffer damage from the abnormal function of the sickle cells and their inability to flow through the small blood vessels correctly. Problems may include:

- Increased bacterial infections (blood, bone, lung)
- Leg ulcers
- Bone / joint damage
- Early gallstones
- Kidney damage and loss of body water in the urine eye damage. (Matthew Heeney, MD, © Children's Hospital Boston, 2011)

1.9 Research problem:

Most of the sickle cell anemia is slight when the condition stable and when the condition worse. This will lead to complications, so ultrasound can detect the splenic size, shape and others.

1.10 Objectives:

1.10.1 General objective:

To detect the prevalence of sickle cell anemia in order to control the other.

1.10.2 Specific objectives:

1. To monitor the spleen size in SCA patient.
2. To assess the reduction of spleen size.
3. To find others associated finding rather than spleen changes.

1.11 Over view of the study:

This study contains about five chapters:

- Chapter One: The introduction.
- Chapter two: The literature review.
- Chapter three: The material and methods.
- Chapter four the results.
- Chapter five discussion conclusion recommendation
- The appendix

Chapter 2

The literature Review

2.1 Anatomy of the spleen

2.1.1 Overview

The spleen is an organ shaped like a shoe that lies relative to the 9th and 11th ribs and is located in the left hypochondrium and partly in the epigastrium. Thus, the spleen is situated between the fundus of the stomach and the diaphragm. The spleen is very vascular and reddish purple in color; its size and weight vary. A healthy spleen is not palpable (CHADBURN, A. 2000) .

2.1.2 Development:-

The spleen develops in the cephalic part of dorsal mesogastrium (from its left layer; during the sixth week of intrauterine life) into a number of nodules that fuse and form a lobulated spleen. Notching of the superior border of the adult spleen is evidence of its multiple origin (see the image below) (CHADBURN, A. 2000) .

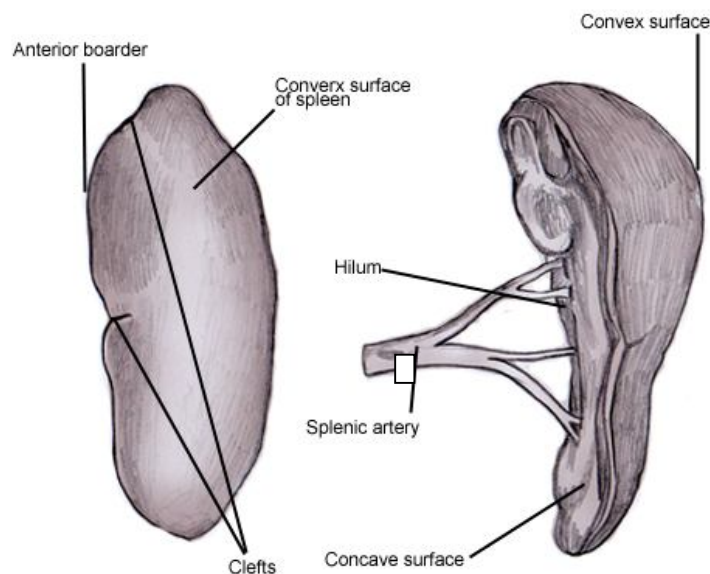


Figure 2-1 Spleen anatomy. The spleen's surfaces and splenic notches

The spleen's two ends are the anterior and posterior end. The anterior end of the spleen is expanded and is more like a border; it is directed forward and downward to reach the midaxillary line. The posterior end is rounded and is directed upward and backward; it rests on the upper pole of the left kidney. (CHADBURN, A. 2000) .The spleen's three borders are the superior, inferior, and intermediate. The superior border of the spleen is notched by the anterior end. The inferior border is rounded. The intermediate border directs toward the right. (CHADBURN, A. 2000) .

The two surfaces of the spleen are the diaphragmatic and visceral. The diaphragmatic surface is smooth and convex, and the visceral surface is irregular and concave and has impressions. The gastric impression is for the fundus of the stomach, which is the largest and most concave impression on the spleen. The renal impression is for the left kidney and lies between the inferior and intermediate borders. The colic impression is for the splenic flexure of the colon; its lower part is related to the phrenicocolic ligament. The pancreatic impression for the tail of the pancreas lies between the hilum and colic impression (see figure 2-2 below) (CHADBURN, A. 2000) .

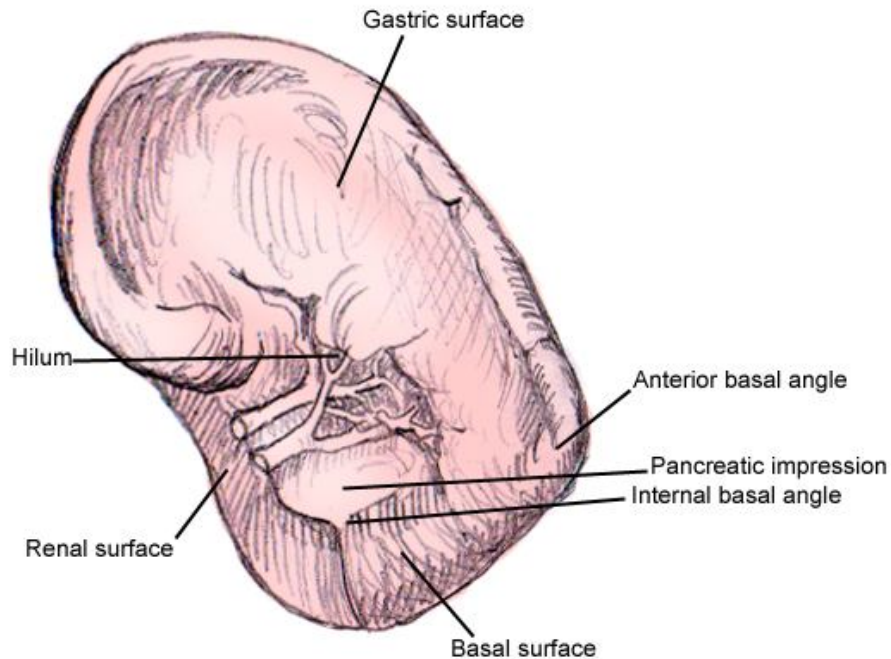


Figure 2-2 shows different surfaces and impressions caused by different organs in relation to the spleen's hilum.

2.1.3 Hilum:

The hilum can be found on the inferomedial part of the gastric impression (see the image above). The hilum transmits the splenic vessels and nerves and provides attachment to the gastrosplenic and splenorenal (lienorenal) ligaments. (CHADBURN, A. 2000) .

2.1.4 Peritoneal relations:-

The spleen is surrounded by peritoneum and is suspended by multiple ligaments, as follows:

- The gastrosplenic ligament extends from the hilum of the spleen to the greater curvature of the stomach; it contains short gastric vessels and associated lymphatics and sympathetic nerves.
- The splenorenal ligament extends from the hilum of the spleen to the anterior surface of the left kidney; it contains the tail of the pancreas and splenic vessels.

- The phrenicocolic ligament is a horizontal fold of peritoneum that extends from the splenic flexure of the colon to the diaphragm along the midaxillary line; it forms the upper end of the left paracolic gutter (CHADBURN, A. 2000) .
- 2.1.4 Visceral relations:-

The visceral surface of the spleen contacts the following organs:

- Anterior surface of the left kidney
- Splenic flexure of the colon
- The fundus of the stomach
- Tail of the pancreas

The diaphragmatic surface is related to the diaphragm; the diaphragm separates the spleen from the pleura and the lung. (CHADBURN, A. 2000) .

2.1.5 Vascular supply:-

The splenic artery supplies blood to the spleen. This artery is the largest branch of the celiac trunk and reaches the spleen's hilum by passing through the splenorenal ligament. It divides into multiple branches at the hilum. It divides into straight vessels called penicillin, ellipsoids, and arterial capillaries in the spleen.(MALINOVSKY,L.,D'ANDREA,V.& ARTICO, M. 1995).

The splenic circulation is adapted for the separation and storage of the red blood cells. The spleen has superior and inferior vascular segments based on the blood supply. The 2 segments are separated by an avascular plane.(MALINOVSKY,L.,D'ANDREA,V.& ARTICO, M. 1995).

Its terminal branches aside, the splenic artery also gives off branches to the pancreas, 5-7 short gastric branches, and the left gastro-omental (gastroepiploic) artery (see figure 2-3)

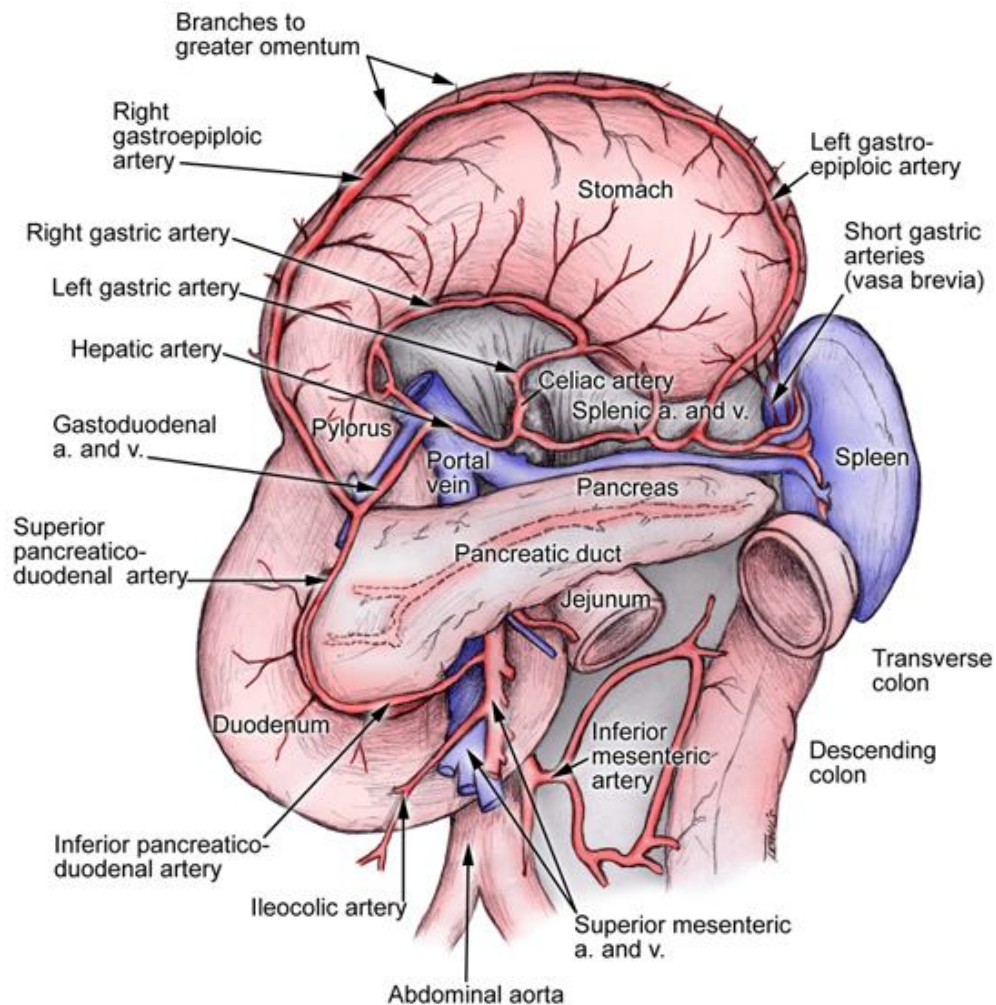


Figure 2-3 Hilum of the spleen along with anatomy of the splenic artery (a) and the splenic vein (v).

2.1.6 Nerve supply:-

Sympathetic fibers are derived from the celiac plexus.

2.1.7 Surface marking:

The spleen is marked on the left side of the back with the long axis of the 10th rib. The upper border is marked along the upper border of the ninth rib; the lower border, along the 11th rib.

The medial end lies 5 cm from the midline. The lateral extension ends at the midaxillary line) (CHADBURN, A. 2000) .

2.1.8 Venous drainage:-

The splenic vein provides the principal venous drainage of the spleen. It runs behind the pancreas (after forming at the hilum) before joining the superior mesenteric vein behind the neck of the pancreas to form the portal vein. The short gastric, left gastro-omental, pancreatic, and inferior mesenteric veins are its tributaries.

(MALINOVSKY, L., D'ANDREA, V. & ARTICO, M. 1995).

2.1.9 Lymphatic drainage:-

Proper splenic tissue has no lymphatics; however, some arise from the capsule and trabeculae and drain to the pancreaticosplenic lymph nodes. (CHADBURN, A. 2000) .

2.1.10 Microscopic Anatomy:-

The spleen is made up of the following 4 components:

- Supporting tissue
- White pulp
- Red pulp
- Vascular system

Supporting tissue is fibroelastic and forms the capsule, coarse trabeculae, and a fine reticulum. The white pulp consists of lymphatic nodules, which are arranged around an eccentric arteriole called the Malpighian corpuscle.

The red pulp is formed by a collection of cells in the interstices of the reticulum, in between the sinusoids. The cell population includes all types of lymphocytes, blood cells, and fixed and free macrophages. The lymphocytes are freely transformed into plasma cells, which can produce large amounts of antibodies and immunoglobulins (see figure 2-4). (CHADBURN, A. 2000) .

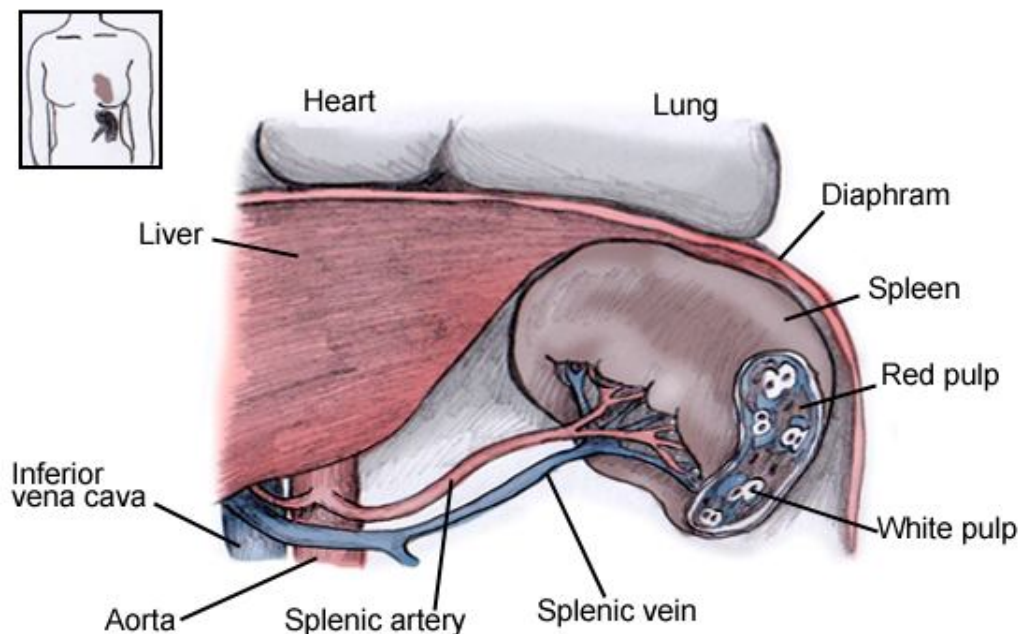


Figure 2-4 shows the spleen's red pulp and the white pulp and its relation to the liver and diaphragm. The vascular system traverses the spleen and permeates it

2.2 Physiology of the spleen:

2.2.1 Immune responses

After antigenic stimulation, increased formation of plasma cells for humoral responses and increased lymphopoiesis for cellular responses occurs. Phagocytosis is one of the spleen's most important functions are phagocytosis. The spleen is a component of the

reticuloendothelial system. The splenic phagocytes include reticular cells, free macrophages of the red pulp, and modified reticular cells of the ellipsoids. Phagocytes in the spleen remove debris, old and effete red blood cells (RBCs), other blood cells, and microorganisms, thereby filtering the blood. Phagocytosis of circulating antigens initiates the humoral and cellular immune responses. (CHADBURN, A. 2000) .

2.2.2 Hematopoiesis

The spleen is an important hematopoietic organ during fetal life; lymphopoiesis continues throughout life. The manufactured lymphocytes take part in immune responses of the body. In the adult spleen, hematopoiesis can restart in certain diseases such as chronic myeloid leukemia and myelosclerosis. (CHADBURN, A. 2000) .

2.2.3 Storage of red blood cells

The RBCs are stored in the spleen. Approximately 8% of the circulating RBCs are present within the spleen; however, this function is seen better in animals than humans.

The spleen is useful for the production of antibodies against antigen present in blood and it is the only organ that performs such functions. Other organs may produce antibodies against antigens seen or present in tissue.

The spleen produces large amounts of B and T lymphocytes through its white pulp. It is the largest site for macrophage aggregations and phagocytic function in the body. It therefore removes old or bad blood cells and platelets. It destroys bacteria and foreign organisms by a process of opsonization and also phagocytosis to include its function of producing antibodies. It stores about 33% of all platelets in the entire body. It is involved in hematopoiesis in the fetus and may be implicated in extramedullary hematopoiesis in certain disease conditions in the adult. The spleen serves in many lower animals and to a little extent in man a reservoir

for storing up blood which may then be released into circulation when it is required, as in sudden loss of blood etc. (CHADBURN, A. 2000) .

2.2.4 Microcirculation in the spleen:

The open system suggests that there is no continuity between the ellipsoids and the sinusoids. Blood from the ellipsoids are discharged into the reticulum of the spleen from where they are absorbed into the sinusoids. (CHADBURN, A. 2000) .

2.2.5 The compromise theory which suggests that both mechanisms take place:

The sinusoids make up the red pulp of the splenic pulp. They are connected to the venules as per any of the mechanisms above and from the venules they discharge their blood into the splenic vein. The sinusoids are lined by special endothelial cells which are banana shaped and contain myofibrils that allow them to contract thereby opening up channels by which blood is discharged into the splenic substance. They are called stave cells. Red blood cell pass through the sinusoidal spaces. When they are old, they are unable to pass across and they are then destroyed by the splenic macrophagic system. (CHADBURN, A. 2000) .

2.3 Pathology of the spleen:

2.3.1 CONGENITAL VARIANTS

2.3.1.1 Accessory spleen

- Accessory spleens are small nodules of splenic tissue which are found in addition to the main bulk of the spleen and are reportedly found in up to 30% of autopsies.²⁰ They arise from failure of the individual clumps of mesenchyme to fuse during the development of the spleen. They may be single and are most commonly found in the splenic hilum, or they may be multiple and located anywhere within the upper abdomen but particularly along the splenic vessels or within the omental layers. Due

to the relationship of the development of the spleen, mesonephros and left gonad, splenic tissue may even be attached to the left ovary or within the scrotum. This is called splenogonadal fusion and is important as it may present as a misrelated to the testis which may result in orchidectomy. Splenunculi vary in size from a few millimeters to a few centimeters and are similar in echogenicity to the splenic parenchyma. A blood supply can be demonstrated on Doppler. They may cause a diagnostic dilemma in that they are often mistaken for lymph nodes in the splenic hilum. . (Rose de Bruyn. 2005) .

2.3.1.2 The wandering spleen

- This refers to a spleen that is highly mobile and either on a long or even absent pedicle. The importance that the spleen may be lying abnormally low within the abdomen or may have an abnormal lie. It has been reported in children with abnormal abdominal musculature such as a sprune belly syndrome.²²As a result of the abnormal pedicle, torsion or twisting of the spleen may occur with resulting compromise of the blood supply to the spleen. In the acute phase of torsion the spleen appears large and hypoechoic because of infarction and congestion ,and in the chronic phase the spleen may be small and difficult to detect. Doppler will demonstrate flow in the spleen. . (Rose de Bruyn. 2005) .

2.3.1.3 SPLENOMEGALY

Causes of splenomegaly include:

- Apparent splenomegaly, due to a displacement of the spleen by a large liver extending across the midline
- portal hypertension

- lymphoma and leukemia; other evidence of disease is usually present, such as hepatomegaly and lymphadenopathy
- infection—viral, bacterial, fungal, protozoal, malaria, mycobacterium and histoplasmosis
- lymphoproliferative disorders
- heart failure and splenic congestion
- infiltrative storage disorders such as Gaucher's disease, mucopolysaccharidosis and Langerhans cell histiocytosis
- hemolytic anemias with extramedullary hemopoiesis—that is, the production of blood cells outside the bone marrow when it is diseased
- focal large splenic lesion due to abscess, cyst or trauma, which may present as splenomegaly
- Extracorporeal membrane oxygenation (ECMO)—this is thought to be related to damage to the red cells, with the spleen enlarging as it removes these damaged cells.

(Rose de Bruyn. 2005) .

2.3.1.4 THE SMALL spleen

This may be caused by:

- Infarction either due to a wandering spleen or conditions such as sickle cell disease
- Congenital absence or hypoplasia of the spleen
- Partial splenectomy
- Celiac disease
- Fanconi anemia. (Rose de Bruyn. 2005)

2.3.1.5 FOCAL SPLENIC LESIONS

2.3.1.5.1 Cysts

Cysts in the spleen are occasionally detected and may be true cysts or pseudocysts. It may be difficult once the cyst is detected to determine the initial etiology, but the wall must be carefully examined to help detect whether it is a primary congenital cyst or a cyst secondary to an infective process or a hematoma; in the latter case, there will be no epithelial lining. However, it is generally not possible to differentiate between true and pseudocysts. (Rose de Bruyn. 2005) .

2.3.15.2 Pseudocysts:

Splenic cysts may rupture or become infected. Epidermoid cysts may also occur in the spleen and may appear as well-defined cystic lesions. There may be calcification within the cyst or within the wall and it may be multiseptated. The cyst in addition may appear complex with internal echoes resulting from hemorrhage or infection .Hydatid infections may also be seen in the spleen, and this generally results from a ruptured liver cyst or systemic infection. They are well defined, single or multiple, with wall calcification. (Rose de Bruyn. 2005) .

2.3.1.5.2 Hemangiomas

Hemangiomas are one of the commonest benign neoplasms in the spleen. The ultrasound appearances of hemangiomas vary according to the size of the blood vessels and may be single or multiple and may have a hyperechoic appearance or may appear cystic if the blood vessels are large. (Rose de Bruyn. 2005) .

2.3.1.5.3 Lymphangiomas

Lymphangiomas in the spleen appear as they do elsewhere in the body, with multiple large cystic spaces filled with lymph. The vascular content is variable, so that some have large

capillaries. Ultrasonically the lesions have multiple cystic spaces with variable thickness and vascularity of the septations. (Rose de Bruyn. 2005) .

2.3.1.5.4 Abscesses

Abscesses in the spleen have similar ultrasound appearances to those in the liver. They are also usually spread by the blood and typically have thick walls and internal echoes. In pediatric practice, children who develop splenic abscesses are those who are immunocompromised. In this instance they are often multiple. Pyogenic abscesses are typically ill-defined hypoechoic lesions on ultrasound and may contain internal debris. These pyogenic abscesses may be hematogenously spread from a primary site of infection such as appendicitis, empyema, osteomyelitis or ear infections. They may be associated with subacute bacterial endocarditis. Fungal abscesses, such as those particularly seen in candida, aspergillus and cryptococcus, are indistinguishable on ultrasound. They have a variable appearance and may appear similar to those seen in the liver such that they may have a well-defined hypoechoic lesion or they may have a bullseye or target appearance. They are typically very small, only a few millimeters, and a linear transducer is preferred when suspected. Usually the spleen and liver are involved together rather than in isolation. (Rose de Bruyn. 2005) .

2.3.1.6 Diffuse splenic abnormalities

2.3.1.6.1 Neoplastic conditions

Both lymphoma and leukemia may involve the spleen in a focal or diffuse manner. When the spleen is diffusely enlarged there may be no focal abnormality with an apparently normal architecture. Lymphoma (both Hodgkin and non-Hodgkin) and leukemia are the commonest neoplastic conditions involving the spleen .Involvement of the spleen may occur in

approximately a third of children with lymphoma. The spleen may be enlarged and may have focal hypoechoic nodules. (Rose de Bruyn. 2005) .

2.3.1.6.2 Storage disorders

Hepatosplenomegaly is seen in all patients with Gaucher disease. This is a congenital disorder where there is lack of the enzyme glucocerebrosidase. Glucocerebrosidase as a result accumulates in the liver and spleen. Gaucher cells may accumulate causing hyper- or hypoechoic nodules. Nodules in the spleen. Extramedullary hemopoiesis, infarction and fibrosis may also occur. (Rose de Bruyn. 2005) .

2.3.1.6.3 Infarction

There are a number of conditions that may cause infarction of the spleen in children, such as sickle cell disease, torsion due to an abnormal pedicle, cardiac emboli, vascular diseases and, rarely, portal hypertension. The spleen is particularly vulnerable to infarction as the splenic artery is an end artery .Ultimately the spleen will appear small with echogenic areas. There may be isolated areas of infarction or the whole spleen may undergo infarction .Patients with sickle cell disease are particularly prone to developing splenic infarcts (Rose de Bruyn2005)

2.3.1.7 SPLENIC TRAUMA

The spleen is one of the most commonly traumatized intra-abdominal organs. Trauma to the spleen may involve:

- Intra splenic parenchymal lacerations.
- Subcapsular hematomas resulting from the parenchymal tear or laceration. (Rose de Bruyn. 2005)

2.4 Previous studies:

There are many studies on SCD around the world. Bakhieta Ibrahim Attalla said that (Abdominal sonographic findings of patients with SCD showed a high incidence of abdominal abnormalities ,especially in solid organs such as the liver, gallbladder ,spleen, and kidney. Repeated vascular occlusion, chronic hemolysis, and anemia contribute to the pathogenesis of multiple abdominal manifestations of SCD). (Bakhieta Ibrahim Attalla, MD 2010) Ma'aji SM, et all that (A total of 71 patients with sickle cell anemia were scanned. The age range was 1-15 years. The commonest TAUS finding in this study was hepatomegaly in 70 (98.6%) patient, followed by splenomegaly in 15 (21.1%), gallbladder wall thickening in 7 (9.7%), and gallbladder stone seen in 4 (5.6%) patients. Autosplenectomy was demonstrated in 3 (4.2%) patients, gallbladder sludge was seen in 2 (2.8%) and increased renal medullary echogenicity was seen in 1 (1.4%) patient.) (Ma'aji SM, et all 2012)

A.A Babadoko, et all that (The mean age of the sickle cell subjects was 23.2 ± 5.3 years, while that of the controls was 22.7 ± 12.4 years. Of the 74 sickle cell subjects, 55.4% were females; while of the 20 controls, 50% were females. Forty one subjects (55.4%) had autosplenectomy and a significant difference existed in the mean splenic size compared with the control ($p < 0.0001$). Only 3 (4.05%) subjects had splenomegaly, while 23 (31%) had a shrunken spleen) (A.A Babadoko, 2012)

Al-Salem AH1 et all that (Only 24 (6.6%) of our patients had autosplenectomy. The splenic index increased with age until about 40 years of age and then gradually decreased, indicating persistence of splenomegaly in our patients into an older age group. Forty-three patients (11.8%) had marked-massive splenomegaly (splenic index $> 120 \text{ cm}^2$) and these had higher HbF levels (mean HbF=22.2%) when compared with those who had autosplenectomy (mean

HbF=14.6). This is significant (P-value=0.0169) and confirms the effect of HbF on persistence of splenomegaly in SCD patients.). (Al-Salem AH1,.1998)

Olatunji AA1, Olatunji PO that (Both mean longitudinal and coronal diameters, and surface area were found to be higher in the sickle cell anemia patients compared to the controls. While the longitudinal diameter increased with age continuously, the coronal diameter decreased after the age of 30 years. There were no significant correlations between the splenic sizes and PCV and number of crises per year) (Olatunji AA1, Olatunji PO. 2001)

Wilson-Okoh DA1, Nwauche CA, Ejele OA. That (The essential splenic change in SCA is splenomegaly and subsequent shrinkage in size (autosplenectomy), which maybe due to several factors. These include: high levels of irreversible sickle cells, decreased HbF associated with increased intravascular sickling and chronic Malaria infection secondary to hyperplasia of the reticulo-endothelial system and increased antibody production especially IgG and IgM. Finally, the clinical complications of these splenic changes such as increased susceptibility to infection, acute splenic sequestration and hypersplenism are also reviewed in this paper.) (Al-Salem AH1,.1998)

Chapter Three

3.1 Material and methods:

This is a prospective study on randomly selection Sudanese children patients with SCD Ultrasonographic studies are do using a real-time gray-scale ultrasound and 3 and 5 Mega Hertz transducer. The patients sample about 35 patient with SCA and 10 children control group.

3.2 Type of the study:

This prospective study deals with the children suffer of SCD that com to SCD clinic.

3.3 Population of the study:-

Children with SCD present to ultrasound department in Mohammed Elamin Hamed Hospital

3.4 Study sample:

The sample size contains 35 patient's aged between 1-16 years with SCD and 10 individuals as control group

3.5 Machine used:

Real time ultrasound Aloka machine with 3.5 and 5 MHz

3.6 Technique for scanning:

Permission is taken from each patient for research purpose. All patients were seated supine a generous couple of gel is applied to the patient for better resolution.

Axial and coronal oblique planes were used to visualize the spleen and other abdominal organs.

The spleen size is recorded plus the splenic vein diameter, other abdominal organs finding were recorded. The collecting data sheet contains, the patient age, gender, spleen size and volume and others abdominal organs. The data of each pt is recorded immediately after finishing the exam. The data was tabulated for analysis and results.

3.7 Data collection:

The data collection from ultrasound investigation and entered data sheet.

3.8 Data analysis:

SSPS program and excel were used to analyzed the data.

3.9 Data storage:

The data was storage on

- Personal computer
- Patient data collection sheet.

Chapter Four

Results

The data for this study was collected from 35 patients of both genders who were affected by sickle cell diseases, the study includes findings of the spleen character especially the length, width, depth (volume).

The study includes the liver length and other abdominal organs.

The data sheets include the patient's age, height, weight for the affected patients with the sickle cell diseases.

The control group (10 individuals) was selected for enriching and comparing with the patients data. The variables with age and gender and comparing with the normal control were also been evaluated.

Table 4.1 Patients age distribution, frequency and percentages

Age classes	Frequency	Percentage %
1-5	14	40%
6-10	10	28.6
11-15	11	31.42
Total	35	100%

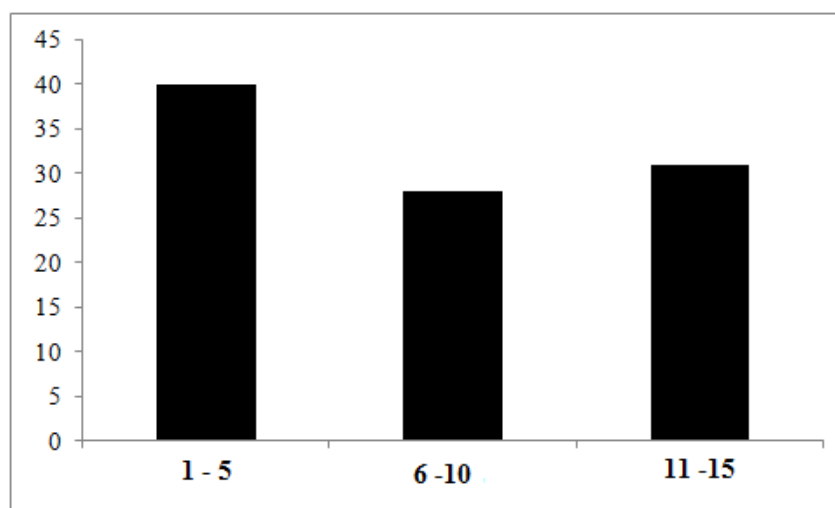


Figure 4-1 Patients age distribution (group A)

Table 4-2 shows the spleen volume of control and SCD patients group (A)

TYPE		N	Mean	Std. Deviation	Std. Error Mean
SPLV	CONTROL	4	33.2925	18.95606	9.47803
	SCA	11	24.8055	13.71849	4.13628

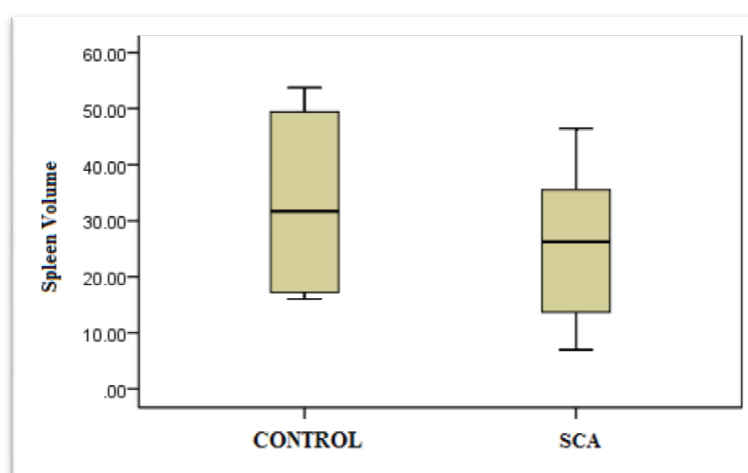


Figure 4-2 shows the value of spleen volume of control and SCD patients (group A)

Table 4-3 shows the liver size of control and SCD patients (group A)

Group Statistics					
TYPE		N	Mean	Std. Deviation	Std. Error Mean
LVS	CONTROL	4	8.3550	1.22563	.61281
	SCA	11	9.4100	1.07703	.32474

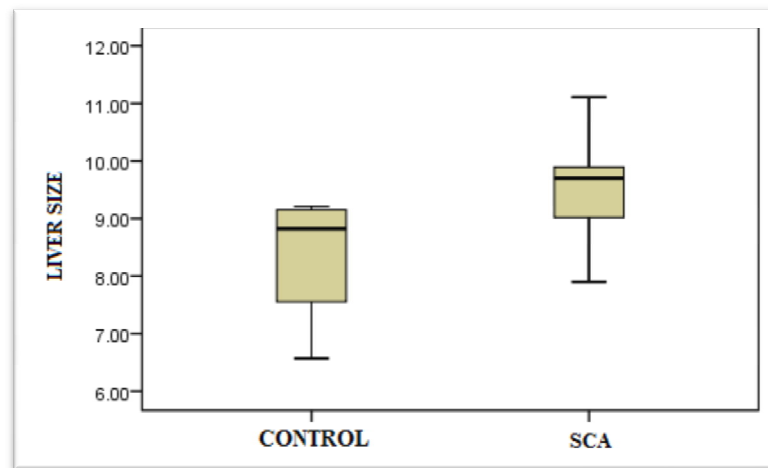


Figure 4-3 shows the value of liver size of control and SCD patients (group A)

Table 4-4 shows the spleen volume of control and SCD patients (group B)

TYPE		N	Mean	Std. Deviation	Std. Error Mean
SPLV	CONTROL	4	81.8150	36.67497	18.33749
	SCA	5	46.3740	51.65244	23.09967

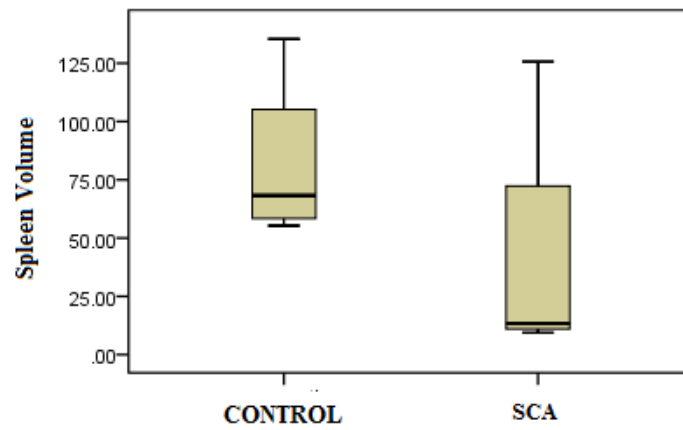


Figure 4-4 shows the value of spleen volume of control and SCD patients(group B)

Table 4-5 shows the liver size of control and SCD patients (group B)

TYPE		N	Mean	Std. Deviation	Std. Error Mean
LVS	CONTROL	4	10.7725	.77569	.38784
	SCA	10	11.4030	1.33786	.42307

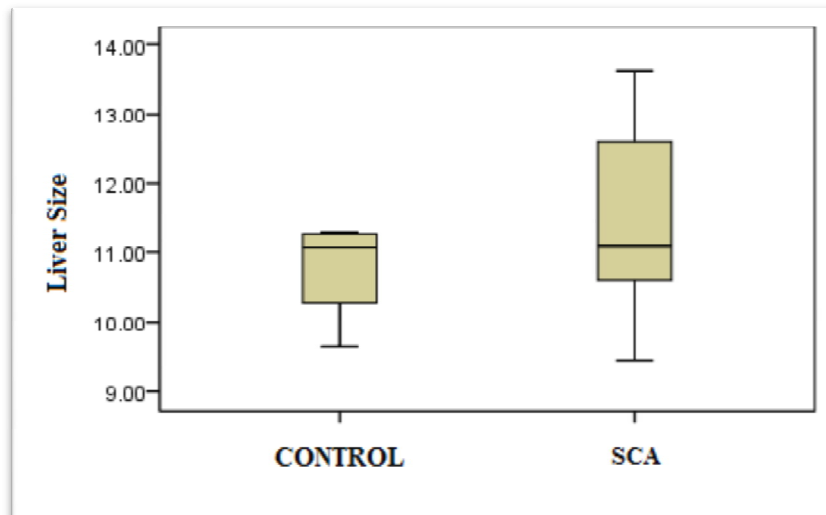


Figure 4-5 shows the value of liver size of control and SCD patients (groupB)

Table 4-6 shows the spleen volume of control and SCD patients (group C)

TYPE	N	Mean	Std. Deviation	Std. Error Mean
SPLV CONTROL	2	110.4950	12.57943	8.89500
SCA	6	29.9600	35.90486	14.65810

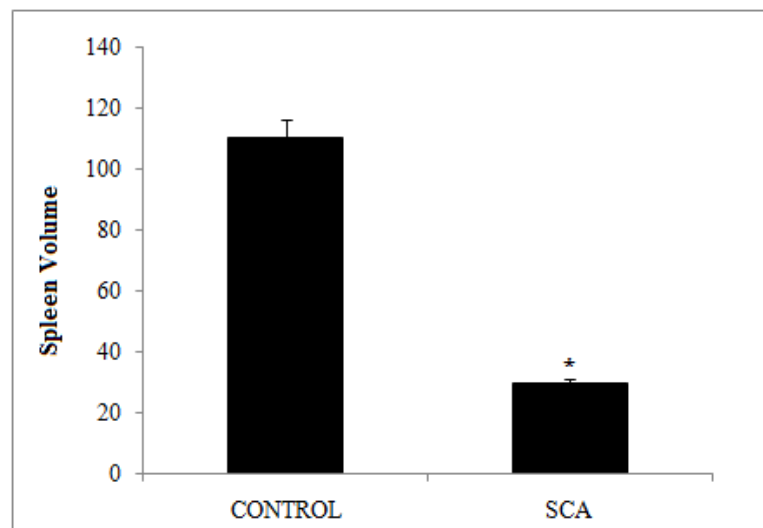


Figure 4-6 shows the value of spleen volume of control and SCD patients (group C)

Table 4-7 shows the liver size of control and SCD patients (group C)

TYPE	N	Mean	Std. Deviation	Std. Error Mean
LVS CONTROL	2	12.3000	.59397	.42000
SCA	11	12.1345	1.54290	.46520

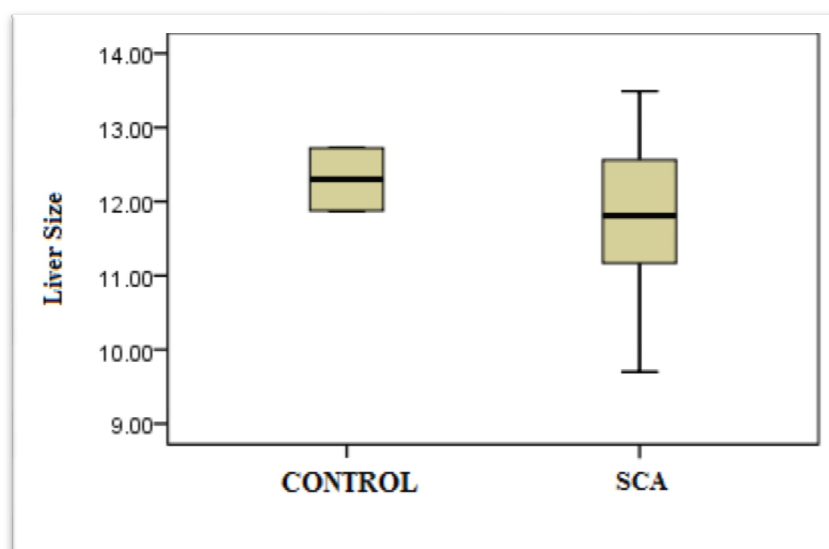


Figure 4-7 shows the value of liver size of control and SCD patients (groupC)

Table 4-8 shows the mean and std. deviation of all groups

Descriptive

	N	Mean	Std. Deviation	Std. Error	95% Confidence Interval for Mean		Minimum	Maximum
					Lower Bound	Upper Bound		
A	13	36.9246	32.21794	8.93565	17.4555	56.3937	6.97	109.60
B	5	46.3740	51.65244	23.09967	-17.7610	110.5090	9.45	125.73
C	6	29.9600	35.90486	14.65810	-7.7198	67.6398	6.57	99.76
Total	24	37.1521	36.30181	7.41008	21.8232	52.4810	6.57	125.73

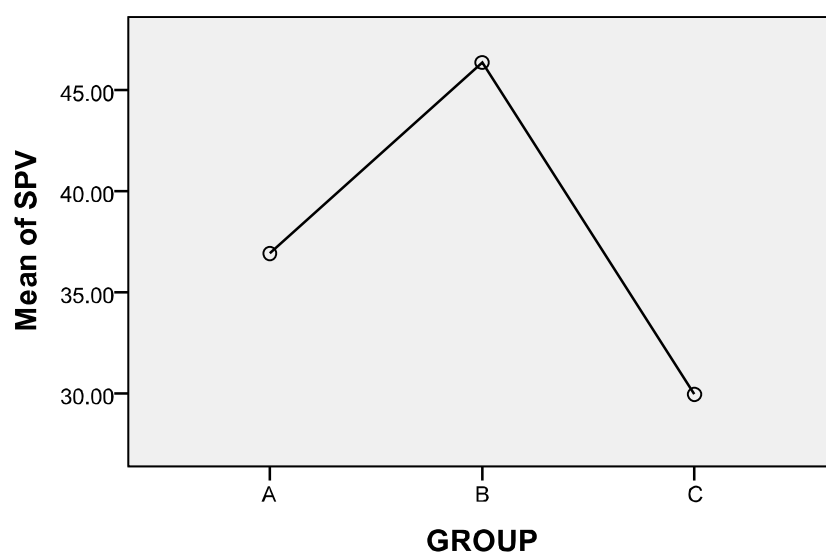


Figure 4-8 shows the relation between groups in mean of spleen volume

Table 4-9 shows the difference of means between all groups

Multiple Comparisons

(I) GROUP	(J) GROUP	Mean Difference (I-J)	Std. Error	Sig.	95% Confidence Interval	
					Lower Bound	Upper Bound
A	B	-9.44938	19.74796	.882	-59.2255	40.3267
	C	6.96462	18.52133	.925	-39.7197	53.6489
B	A	9.44938	19.74796	.882	-40.3267	59.2255
	C	16.41400	22.72367	.753	-40.8626	73.6906
C	A	-6.96462	18.52133	.925	-53.6489	39.7197
	B	-16.41400	22.72367	.753	-73.6906	40.8626

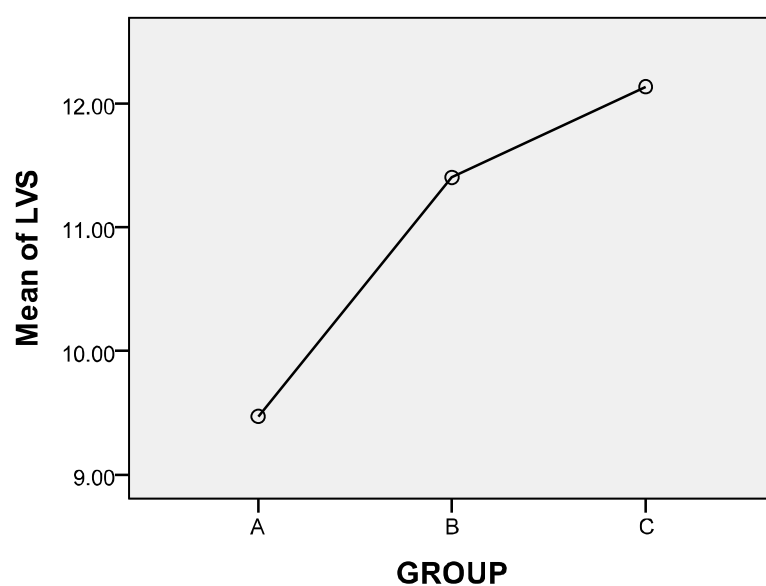


Figure 4-9 shows the comparison of liver size mean between all groups

Table 4-10 shows the relation between groups in mean of liver size

	N	Mean	Std. Deviation	Std. Error	95% Confidence Interval for Mean		Minimum	Maximum
					Lower Bound	Upper Bound		
A	14	9.4707	1.06955	.28585	8.8532	10.0883	7.43	11.11
B	10	11.4030	1.33786	.42307	10.4460	12.3600	9.45	13.63
C	11	12.1345	1.54290	.46520	11.0980	13.1711	9.70	15.46
Total	35	10.8600	1.73735	.29367	10.2632	11.4568	7.43	15.46

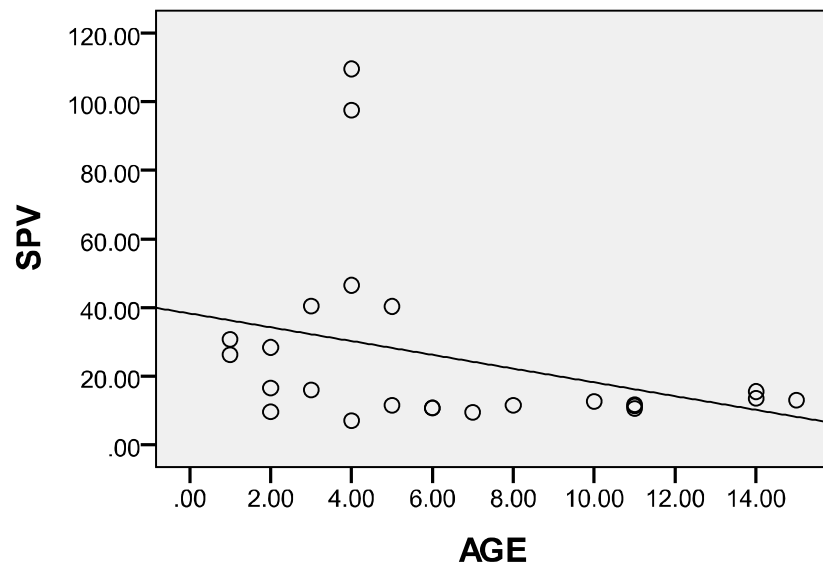


Figure 4-10 shows the relation between spleen volume and age

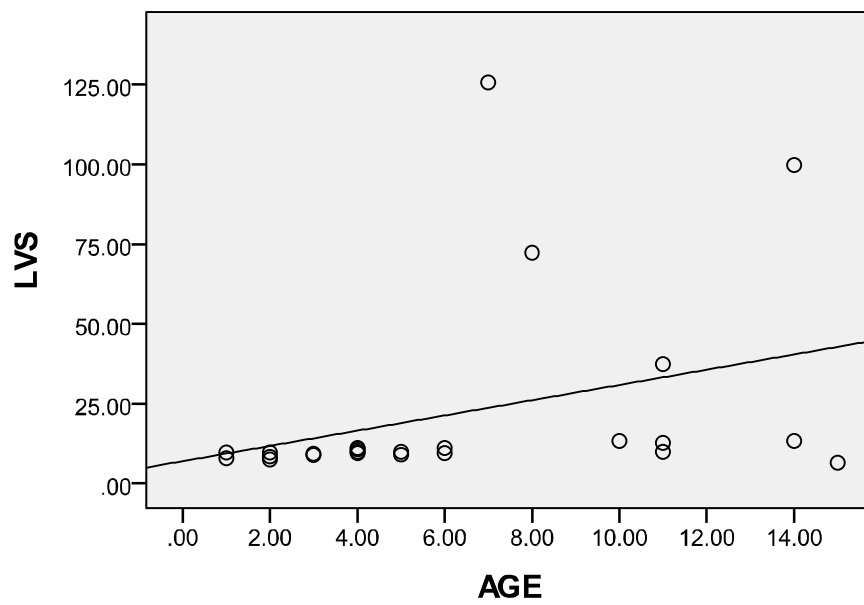


Figure 4-11 shows the relation between liver size and age

Table 4-12 shows the comparison of liver size mean between all groups

(I) GROUP	(J) GROUP	Mean Difference (I-J)	Std. Error	Sig.	95% Confidence Interval	
					Lower Bound	Upper Bound
A	B	-1.93229*	.54175	.003	-3.2636	-.6010
	C	-2.66383*	.52719	.000	-3.9593	-1.3683
B	A	1.93229*	.54175	.003	.6010	3.2636
	C	-.73155	.57170	.417	-2.1364	.6733
C	A	2.66383*	.52719	.000	1.3683	3.9593
	B	.73155	.57170	.417	-.6733	2.1364

*. The mean difference is significant at the 0.05 level.

Table 4-13 shows the correlation between age and spleen volume

Correlations			
		AGE	SPV
AGE	Pearson Correlation	1	-.329
	Sig. (2-tailed)		.116
	N	24	24
SPV	Pearson Correlation	-.329	1
	Sig. (2-tailed)	.116	
	N	24	24

Table 4-14 shows the correlation between age and liver size

Correlations		AGE	LVS
AGE	Pearson Correlation	1	.332
	Sig. (2-tailed)		.113
	N	24	24
LVS	Pearson Correlation	.332	1
	Sig. (2-tailed)	.113	
	N	24	24

Table 4.15 Ultrasound Findings in patients with sickle cell disease

	RT Kidney Texture	LT Kidney Texture	Gall Bladder	Liver Texture	Spleen Texture
Normal	34 (97.1%)	34 (97.1%)	33(94.3%)	34 (97.1%)	34 (97.1%)
Findings	Increased echogenicity 1(2.9%)	Increased echogenicity 1(2.9%)	(Stone) Increased echogenicity 2(5.7%)	(Cyst) Decreased echogenicity 1(2.9%)	Increased echogenicity 1(2.9%)

Chapter Five

5.1 Discussion

The main objective of this study was to evaluate the prevalence of SCA and their effects on abdominal organs .The data was collected from 35 patients their ages between 1 year to 15 years old and 10 normal children as control group. Ultrasound scans were done in Mohammed El amine Hamid of children's hospital at ultrasound clinic. Spleen volume and liver size were measured in all data.

The result of this study showed that the mean value of SCD patients of group A (1-5) years old is (33.29) . This value is slightly smaller than the spleen volume for control group (24.8) (table 4-2).

For the same group from table 4-3 comparing the liver size mean of SCD pt(8.35) and control group(9.41) we noted there is small difference.

For group B (6-10) years old also the result revealed there was difference between the spleen volume mean values of SCD patients (81.81) comparing with control group (46.37) table (4-4) and figure (4-4).

For the same group from table (4-5) and figure (4-5) also there was small difference between liver size mean values of patient comparing to control group.

The results of last group C (11-15) years old similar for other groups but in this group the difference of SCD patient spleen volume values comparing with control group was very clear table (4-6) and figure (4-6) .

The above results was consistent with previous studies which stated that there is a high incidence of abdominal abnormalities, especially in solid organs such as the liver, gallbladder ,spleen, and kidney) (Bakhieta Ibrahim Attalla, MD 2010) ,(A.A Babadoko, 2012), (Wilson-Okoh DA1, Nwauche CA, Ejele OA. 2006) .

Our result showed that there is no significant correlation but there is negative relationship between spleen volume and age (duration of disease), (see table 4-13 and figure 4-10) .

On the other hand there is no significant correlation, however there is positive correlation between liver size and age. see table 4-14 and figure 4-11

The spleen consider as a target organ affected by SCD for its important role in circulation of blood . Splenic sequestration crisis (SSC) occurs when the sickled red blood cells become trapped inside the spleen. This causes the spleen to swell and at the same time leads to a dangerous decrease in the number of red blood cells in the rest of the body. After repeated episodes of splenic sequestration, the spleen becomes scarred, and permanently damaged. Most children, by age 8, do not have a functioning spleen either from surgical removal, or from repeated episodes of splenic sequestration.

From table 4-15 we show there was about one patient have renal changes (increased echogenicity) ,two patient had gall stone , this last tow results consistent with previous study of(Ma'aji SM, et all), one patient have hepatic cyst and one patient have splenic texture changes (increased echogenicity) and also have a jaundie.

During this study we found that about five patient have not spleen (autosplenectomy) from group B ,and also from group C also we show five patient have autosplenectomy, this result consistent with previous study of(Ma'aji SM, et all 2012), (A.A Babadoko, 2012), (Al-Salem AH1,.1998),and (Al-Salem AH1,.1998).

5.2 Conclusion

Most of the sickle cell anemia is slight when the condition stable and when the condition worse. This will lead to complications, so ultrasound can detect the splenic size, shape and others.

By the end of this study found that the ultrasound examination is best choice for detection and follow up the SCD and their complications.

The spleen is the most organ which affected by (SCD) followed by liver, gallbladder and kidneys'.

5-3Recommendation

- An investigation before marriage for affected tribes members should be performed.
- Early investigations for newborns in affected tribes must be encouraged.
- A permanent clinics in affected areas should be established
- Regular examination especially ultrasound for patient as follow up should be done.

References

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Appendix

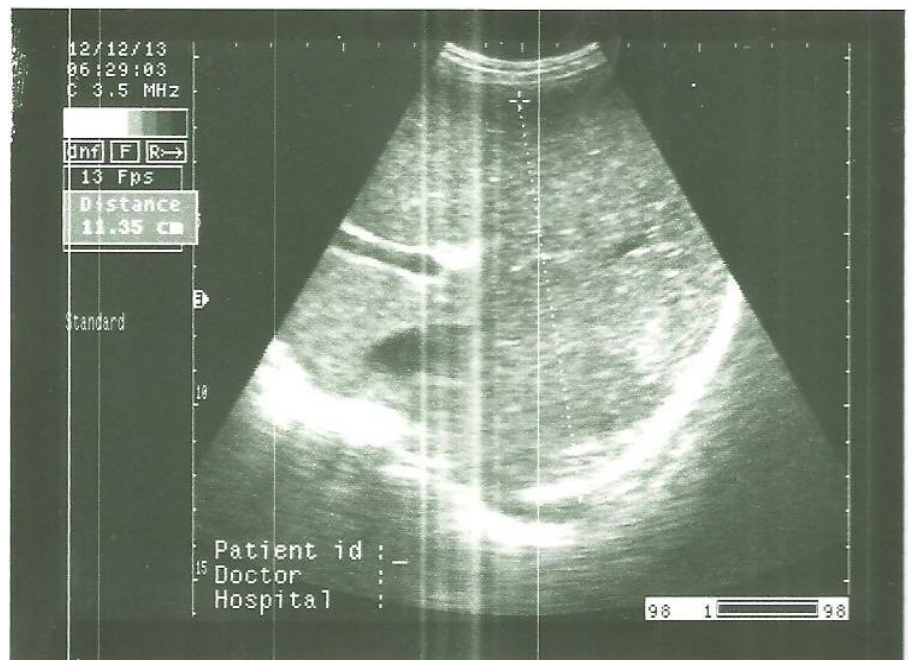


Image explain normal liver size

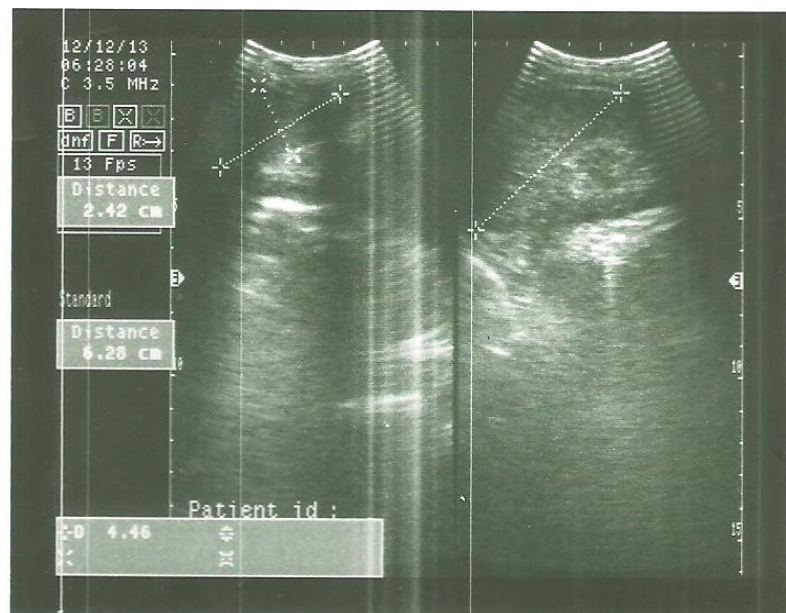


Image explain measurement of spleen (length,depth,width)

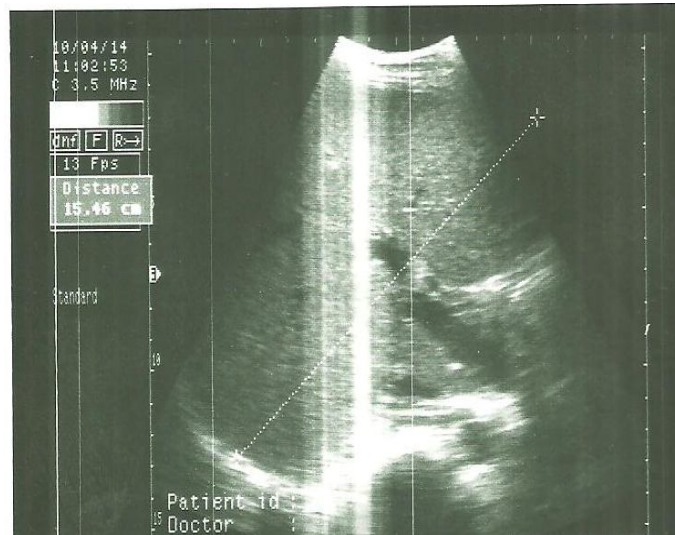


Image explain hepatomegally

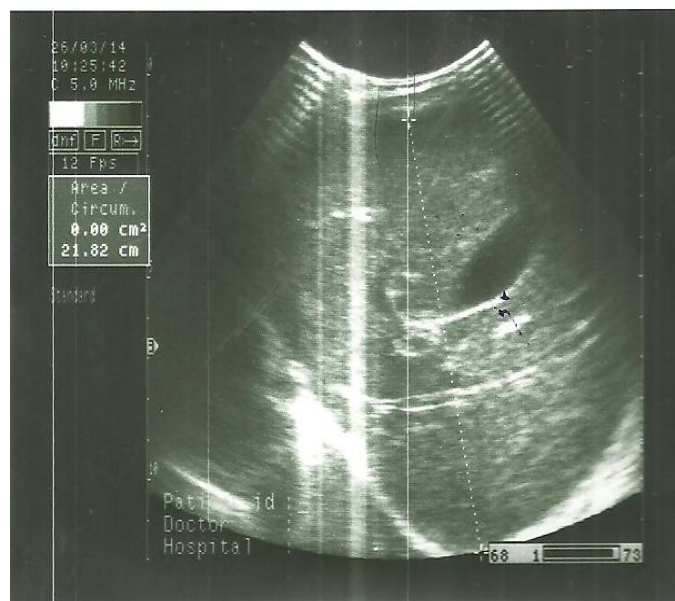


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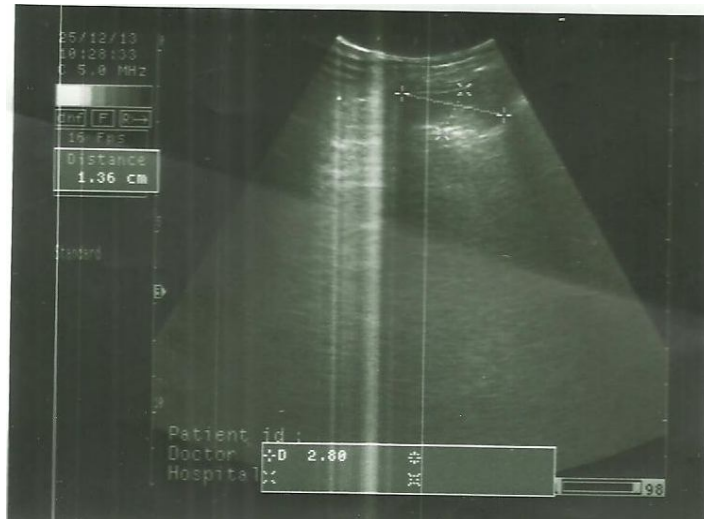


Image explain small spleen

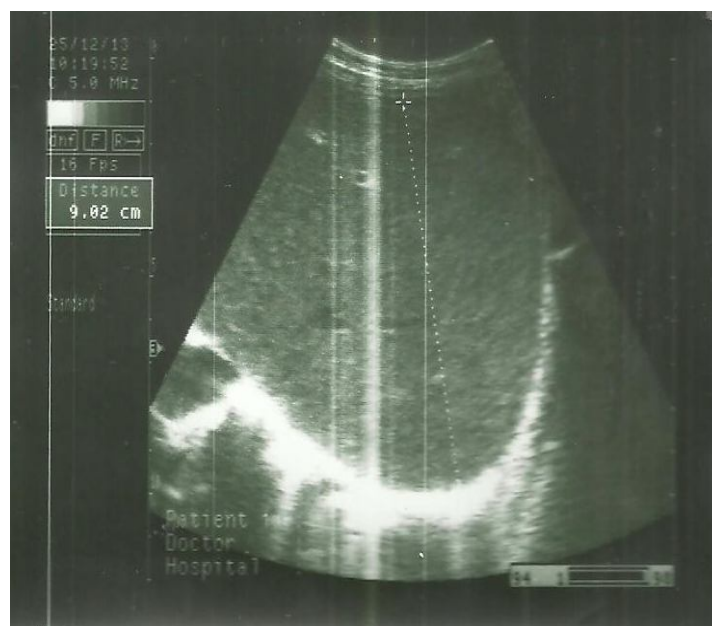


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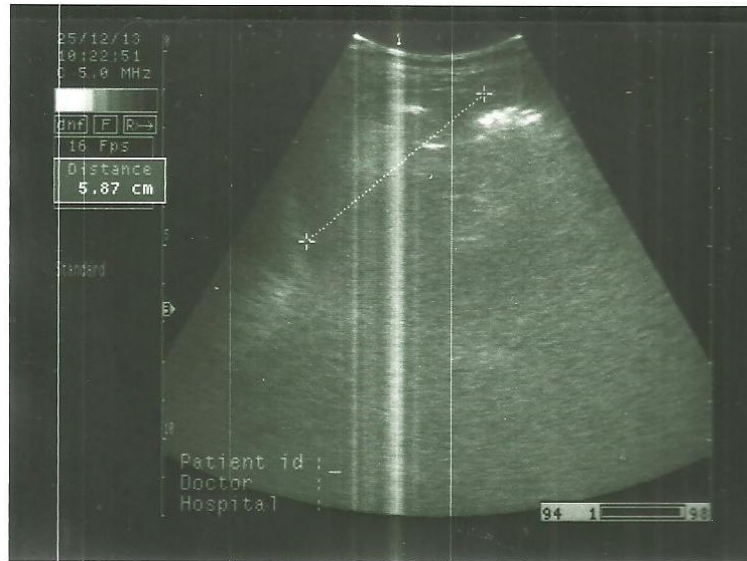


Image explain small spleen

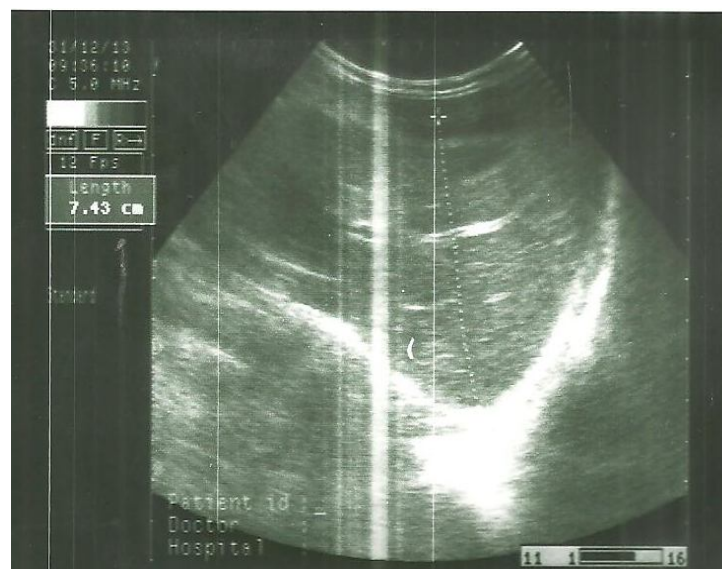


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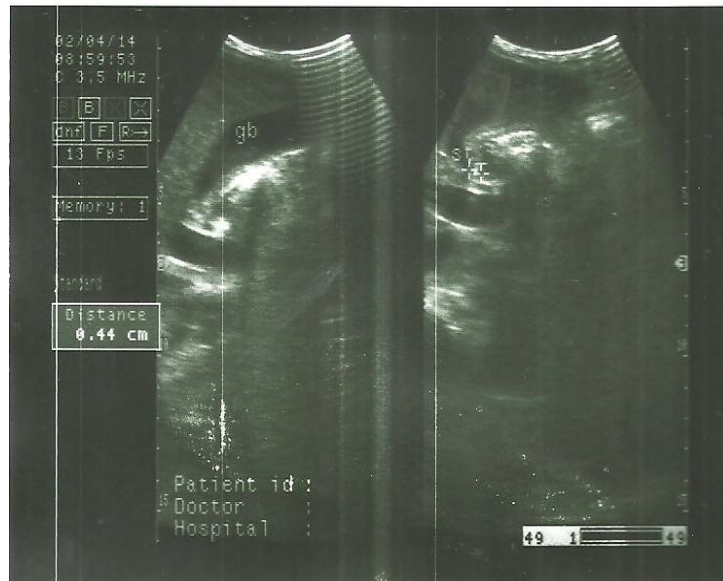


Image explain gall stone

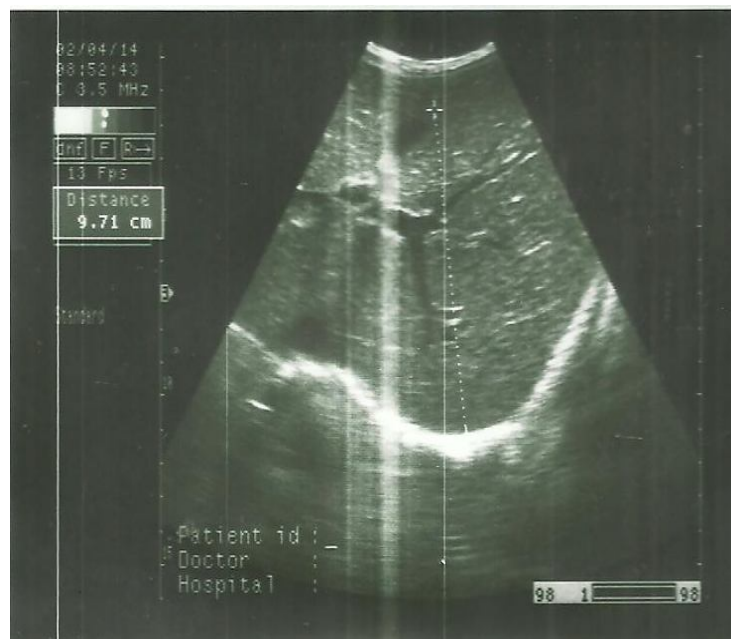


Image explain normal liver

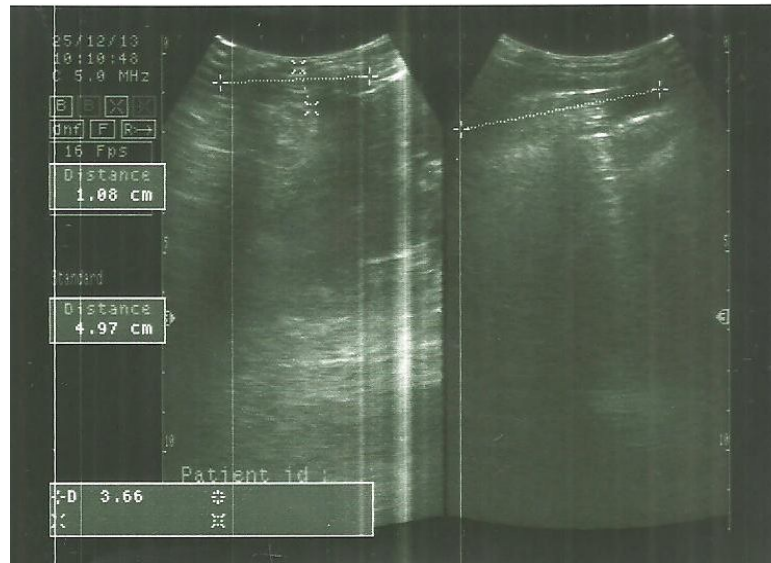


Image explain normal spleen measurement

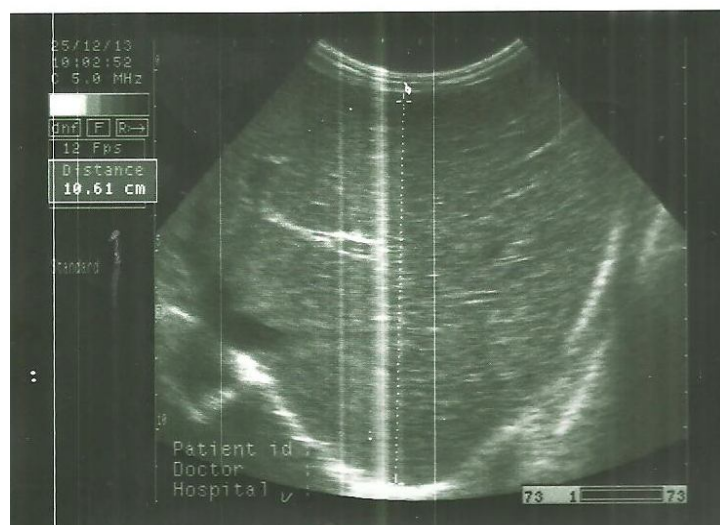


Image explain normal liver size

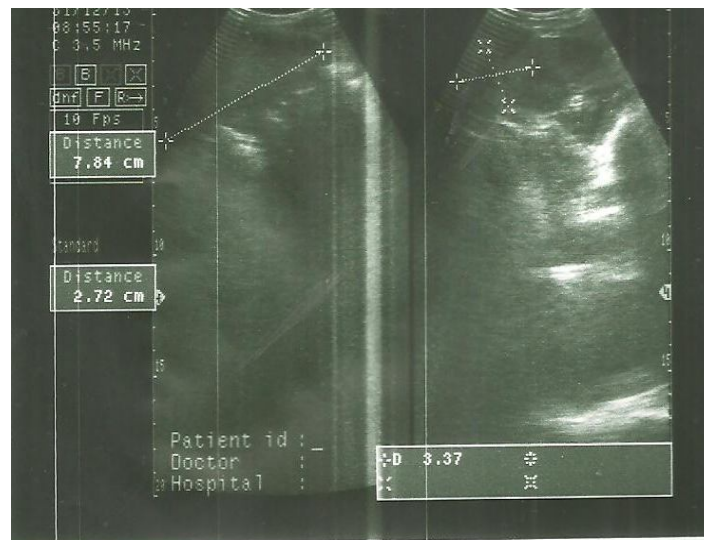


Image explain normal spleen

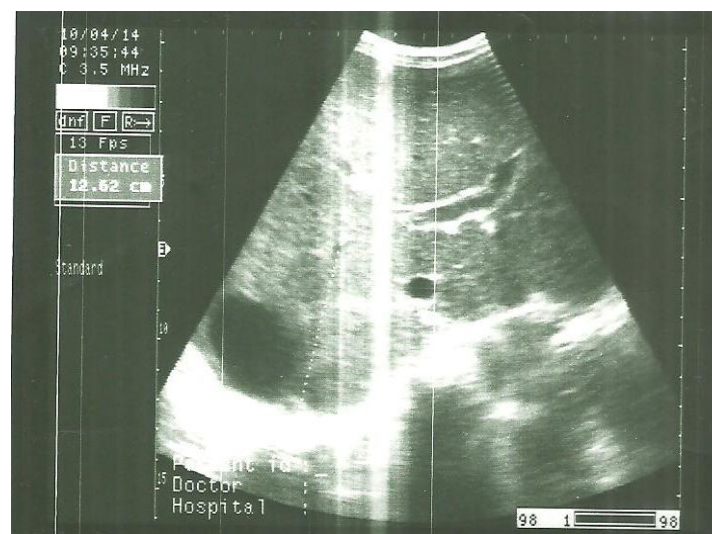


Image explain liver cyst

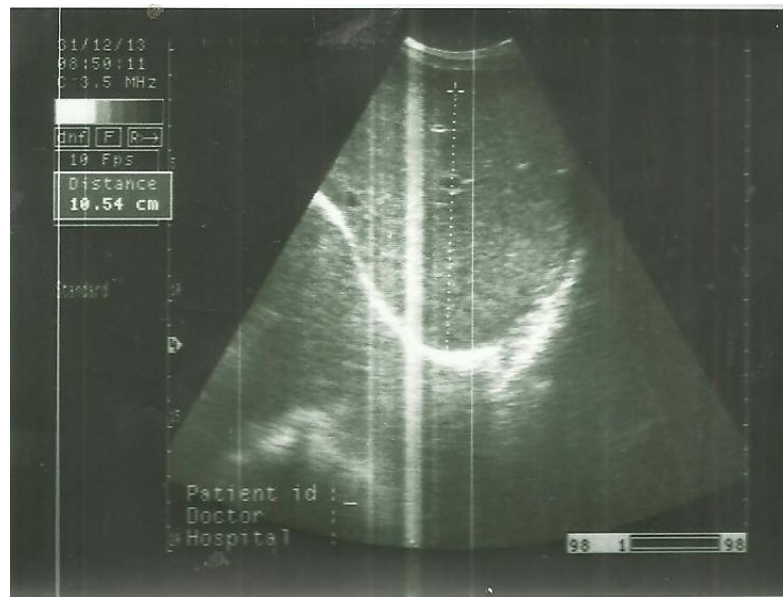


Image explain normal liver size

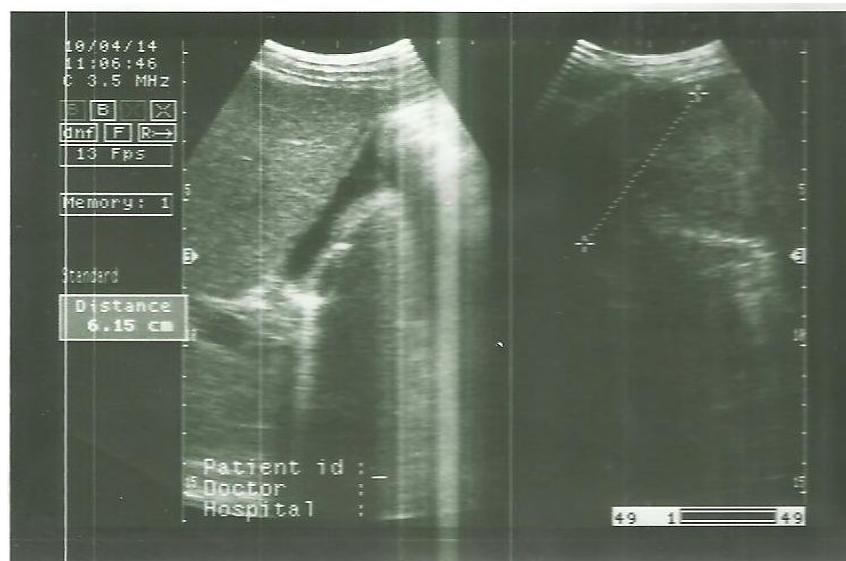


Image explain increased spleen echotexture

