

## Quality Of Life of Children with Sick Cell Anemia

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**Abstract ;Background:** Sick Cell Anemia (SCA) Is One Of The Most Common Chronic Inherited Diseases Worldwide. It Results From Destruction Of An Abnormal Shaped Red Blood Cells That Known As Sickling .This Shape Disrupts The Normal Flow Of Rbcs Through The Blood Vessels Of The Body Which Deprive The Prevent Tissues From Receiving Adequate Oxygen. It Affects Many Aspects Of The QOL Including Physical, Psychological, Intellectual, And Social Aspects Of School-Age Children. A Competent And Qualified Nurse Has A Comprehensive Role In Assessing And Maintaining QOL Of Children With SCA. **Objective:** The Aim Of This Study Was To Assess The QOL Of Children With Sick Cell Anemia **Setting:** This Study Was Conducted In Out And In Patient Departments At Al-Diwaniya Pediatrics Teaching Hospital-Iraq. **Subjects:** Convenience Sample Of 100 Children With Sick Cell Anemia And Their Mothers Comprised The Study Subjects And Who Fulfill The Following Criteria, A- Age Ranges From 6 To12 Years. B- Free From Any Associated Diseases. **Tools:** Three Tools Were Used To Collect The Needed Data, Tool I Namely; Assessment Of Biosociodemographic And Medical Data Of Children With Sick Cell Anemia, **Tool II** Namely; Assessment Of Quality Of Life Of Children With Sick Cell Anemia Structured Interview Schedule, **Tool III** Namely; Visual Analogue Scales (VAS). **Results:** It Was Illustrated That There Were Positive Correlations And Significant Associations Between The Children's Reports And Their Level Of Quality Of Life In Most Categories; Dietary Habit, Rest And Sleep, Intellectual Aspect, Social State, Communication Skills And Dealing Approach Of Disease Symptoms And Treatment Process[(R= 0.931\*, P= <0.001\*), (R=0.931\*, P=<0.001\*), (R=0.495\*, P=<0.001\*), (R= 0.263\*, P=0.008\*), (R= 0.972\*, P=<0.001\*), (R=0.963\*, P=<0.001\*), (R=0.295\*, P=0.003\*) Respectively]. While, Correlation Were Not Found In Two Categories Only [Emotional Aspect And Pain Experienced] Where All Children Have Been Plotted In Poor Level (100%). **Conclusion:** Based On The Findings Of The Current Study, It Is Concluded That The SCA Is Greatly Affecting The Children's Quality Of Life. Whereas, It Was Found That More Than Two Third Of Studied Children Had Poor Quality Of Life And Neutral For Less Than Quarter Of Them. While, Small Percentage Of The Studied Children Had Good Quality Of Life.

**Key Words:** Quality Of Life; Children; Sick Cell Anemia; Children With Sick Cell Anemia.

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### I. Introduction

Sickle Cell Anemia (SCA) Is Inherited Red Blood Cell Disorders. Children With SCA Have Abnormal Hemoglobin, Called Hemoglobin S Or Sick Hemoglobin In Their Red Blood Cells. They Have Abnormal Hemoglobin Genes, One From Each Parent. In All Forms Of SCA, At Least One Of The Two Abnormal Genes Causes A Person's Body To Make Hemoglobin S. This Is The Most Common And Often Most Severe Kind Of Sick Cell Disease<sup>(1)</sup>.

Internationally, According To World Health Organization(WHO), (2011)<sup>(2)</sup>, It Is Estimated That Each Year Over 300, 000 Children With Severe Forms Of SCA Are Born. The Majority Of Them Are In Low And Middle Income Countries, About 2000 Children Born With SCA Each Year In The United States<sup>(3)</sup>. In Egypt At (2015)<sup>(4)</sup>, The Incidence Of Sick Cell Anemia Is 2,238 Children. Whereas, In Iraq The Incidence Is 25,375 Children At 2015<sup>(4)</sup>.

Sickle Cell Anemia Is Caused By Homozygosis For The Sick Gene, The Rbcs Have An Abnormal Crescent Sick Shape That Makes Them Sticky And Rigid. The Loss Of Red Blood Cell Elasticity Is Central To The Pathophysiology Of SCA. Normal Rbcs Are Quite Elastic, Which Allows The Cells To Deform And Pass Through Capillaries. In SCA, Low Oxygen Tension Promotes Red Blood Cells Sickling And Repeated Episodes Of Sickling Damage The Cell Membrane And Decrease The Cells Elasticity. These Cells Fail To Return To Normal Shape When Normal Oxygen Tension Is Restored. As A Consequence, These Rigid Blood



Cells Are Passing Through Narrow Capillaries Leading To Vessel Occlusion And Ischemia. The Actual Anemia Of The Illness Is Caused By Hemolysis And Destruction Of The Red Cells Because Of Their Shape. Although The Bone Marrow Attempts To Compensate By Creating New Red Cells, It Does Not Match The Rate Of Destruction, Healthy Rbcs Typically Function For 90–120 Days, But In SCA Only Last For 10–20 Days<sup>(5)</sup>.

There Are Many Manifestations Of SCA Usually Show Up At A Young Age. It Is Generally Occur Around The 6-Months, They Include Irritability, Fussiness In Children, Jaundice, Swelling And Pain In Hands And Feet, Frequent Infections And Chest Pain. Physical Findings Are Normal At Birth And Symptoms Are Unusual Seen Before Age 3-4 Months Because High Level Of Fetal Hemoglobin Inhibit Sickling. Moderately Severe Hemolytic Anemia May Present By The Of Age 1 Year. Recurrent Episode Of Vaso-Occlusive Crisis And Tissue Ischemia Cause Acute And Chronic Problems. Moreover, Recurrent Episodes Of Abdominal And Musculoskeletal Pain May Occur Throughout Life<sup>(6,7)</sup>.

Therapeutic Management Of Children With SCA Include Antibiotics, High Fluid Intake, Folic Acid Supplementation, Hot Application On Sites As Well As Analgesics. Pneumococcal And Meningococcal Vaccination Are Recommended. Multiple Transfusion Of Packed Red Blood Cell May Be Required To Maintain Adequate Hemoglobin Level. Children In Severe Pain May Need Continuous Intravenous Infusion Containing A Narcotic. Currently, Hematopoietic Stem Cell Transplantation Has A Curative Effect For SCA. Moreover, Prevention Of Both Infection And Dehydration Have Crucial Impact On The Prevention Of Vaso-Occlusive Crisis<sup>(8)</sup>.

The World Health Organization Defines QOL As Individual Perception Of Their Position In Life In The Context Of The Culture And Value System In Which They Live And Relation To Their Goals, Expectation Standard And Concern<sup>(9)</sup>. Quality Of Life Concept Is Affected By The Persons' Physical, Psychological State, Level Of Independence, Social Relationship And Environment. Additionally, SCA Could Affect Quality Of Children's Life From All Dimensions. Physically Includes Diet, Physical Activity, Rest And Sleep, Management Of Pain, Fever, And Vaso-Occlusive Crisis. Besides, Emotional Condition Of Children Could Also Be Affected Due To Body Image Disturbance, Fear, Anxiety About The Need For Future Treatment Can Interfere With Daily Living. Intellectual Aspect Also Can Be Affected As A Result Of Learning Difficulties<sup>(10)</sup>.

The Pediatric Nurse Has A Multiple Roles, One Of The Most Crucial Role Is The Assessment Phase Of Children's Quality Of Life Who Are Suffer From SCA. Assessment Include Disease Progression And Consequence Of The SCA As Well As Its Impact On Children's Life During School Age Period. Adequate And Proper Assessment Of The QOL Reflected On Their Sickle Cell Anemia Condition Will Help The Pediatric Nurse To Develop A Plan Of Care For Encouraging Children To Achieve Better Quality Of Life That Will Reflect On Their Health<sup>(11,12)</sup>.

#### **Aim Of The Study Was:**

This Study Aims To Assess The Quality Of Life Of Children With Sickle Cell Anemia.

#### **Research Question:**

What Is The Quality Of Life Of Children Who Are Suffering From Sickle Cell Anemia?

## **II. Materials And Method**

#### **Research Design:**

Descriptive Research Design Was Used In The Present Study.

#### **Setting:**

This Study Was Conducted In Out And In Patient Departments At Al-Diwaniya Pediatrics Teaching Hospital-Iraq.

#### **Subjects:**

A Convenience Sample Of 100 Children With Sickle Cell Anemia And Their Mothers Comprised The Study Subjects And Fulfill The Following Criteria:

- 1- Age Ranges From 6 To 12 Years.
- 2- Free From Any Associated Diseases.

#### **Tools For Data Collection:**

Three Tools Were Used To Collect The Necessary Data For This Study.

**Tool I: Assessment Of Biosociodemographic And Medical Data Of Children With Sickle Cell Anemia;** This Tool Was Developed By The Researcher After Reviewing The Relevant And Recent Literature<sup>(6,8,13)</sup>. It Was Included The Demographic Data Of Children; Age, Gender, Level Of Education, Birth Order And Residence. Mothers Demographic Data; Age, Level Of Education And Occupation. The Children's Clinical Data Comprised The Onset, Duration And Type Of Treatment Of Sickle Cell Anemia Such As Blood Transfusion: Frequency And Side Effect Of Blood Transfusion.



**Tool II: Assessment Of Quality Of Life Of Children With Sickle Cell Anemia Structured Interview Schedule:**

This Tool Was Developed By The Researcher After Thorough Review Of Related And Recent Literature<sup>(10,13,14)</sup> And Guided By Varni Et Al. (2011)<sup>(15)</sup> To Assess The Quality Of Life Of Children With Sickle Cell Anemia, It Included:

- Physical State Of Children Related To The Disease Process Including:
  - Dietary Habits As Adequate Intake Of Iron, Folic Acid, Vitamin C, Protein, Copper And Cobalt, Water, Water With Fever And Exercise. Avoiding Of Iron And Vitamin C Intake With Frequent Blood Transfusion And Avoiding Of Taking Cold Drinks.
  - Physical Activities, Such As Feeling Tired During Routine Home Activities, Personal Hygiene And Bathing, Running Of Short Distance, Walking Of Long Distance, Performing Exercise And Avoiding Physical Activities In Cold Weather.
  - Rest And Sleeping Pattern Such As Enough Sleeping During Night, Sleeping Interruption During Night Related To Diseases, Taking Enough Rest At Daytime And Taking Warm Bath When Physically Tired.
- Intellectual Aspect Such As School Including The Findings Difficulties In Concentration During School Day, In Doing School Duty, Frequent Absent From School Due To Disease And When Doing The Daily Study.
- Emotional State As Children's Feeling Related To The Disease Including; Fear, Sad, Loneliness Most Of The Time, Lack Of Confidence In Personal Abilities, Worry That To Be Away From Home And Uncomfortable And Unsatisfied In Daily Performance.
- Social State Such As Staying Alone Most Of Time, Child's Inability To Do Things That The Child's Age Can Do And The Difficulty In Participating Playing With Children.
- Communication Problem When Have Pain Including; The Disease Condition That Burden The Communication Such As Telling Others When Feel Pain, Telling Doctors And Nurses About What Feel, Understanding Others To What Extent Feel Pain And Telling Others That Having SCA.
- Children's Dealing Approach With The Disease Symptoms Such As Using Comfortable Measures As Stay In Calm Place And Keeping Away From Light, Taking Some Drinks As Ginger, Mint, Lemon And Taking Of Analgesic In Case Of Headache, Using Wet Cloth (Tipped Compresses), Hospital Admission Or Taking Medication In Fever, Avoiding Of The Tight Clothing And/Or Cold Application And Following Doctor's Orders In Arms And Legs Swellings, Using Of Non-Pharmacological Agents As Distraction, Relaxation And Massage And Taking Medication According To Doctor's Order In Bone And Joint Pain.
- The Experienced Pain Intensity As Mild, Moderate And Severe, The Sites Of Feeling Pain As Whole Body, Arms, Legs, Chest, Back And Stomach.
- The Treatment Process Such As Compliance With Taking Medication, Taking Analgesic When They Feel Pain And Feeling Worry When Received Blood Transfusion.

The Children And Their Mothers Responses To These Statements Were Done On A Three Likert Scale That Ranges From 1-3 As Following; Never =1, Sometimes = 2, Often = 3.

According, The Total Score Of Quality Of Life Of Children With SCA Was 80. It Has Been Distributed As Follows:

- Poor = Less Than 60%.
- Neutral = 60% To Less Than 65%.
- Good = 65% And More.

**Tool III: Visual Analogue Scales (VAS) :**

This Tool Was Developed By Cline Et Al. (1992)<sup>(16)</sup>. It Is A Self-Reported Device That Is Used To Report Subjective Data Concerning Pain. It Is A Verbal Descriptive Scale Consists Of A 10 Cm Straight Line Which Represents A Continuum Of Intensity; 0 = No Pain, 1 To Less Than 4 = Mild Pain, 4 To Less Than 7 = Moderate Pain, 7 To Less Than 10 = Severe Pain, 10 = Unbearable. It Was Adopted To Measure The Severity Of Pain In This Study.

**Method**

- Official Letter From The Faculty Of Nursing, Alexandria University Was Directed To The Responsible Authorities Of Outpatient Department, University Children's Hospital At Al-Diwanay Pediatric Hospital To Get Their Permission For Conducting This Study After Explaining Its Purpose.
- Tool I And II Were Developed By The Researcher And Tested For Their Content Validity By Five Experts In The Pediatric Nursing Field. The Validity Was 87.5% For Tool I And 98.4 For Tool II.
- Reliability For The Tool I And II Was Ascertained Using The Cronbach's Coefficient Alpha Test. The Reliability For Tools I Was  $\alpha=0.87$  And For Tool II Was 0.76
- A Pilot Study Was Carried Out On 10% Of Children With SCA And Their Mothers In Order To Check And Ensure Its Clarity And Feasibility. Those Children And Their Mothers Were Excluded From The Subjects.



- Every Child And His Mother Were Interviewed Individually For 30-40 Minutes During Hospitalization In The Medical Ward.
- Data Were Collected Over 4 Weeks Period Starting From November Till December 2017.

**Ethical Considerations:**

- Oral Informed Consent Was Obtained From All Mothers After Providing Appropriate Explanation About The Purpose Of The Study.
- Privacy And Anonymity Of The Participant Were Assured.
- Confidentiality Of Data Was Assured.
- The Children And Their Mothers Have Right To Withdraw From The Study At Any Time.

#### **IV. Indentations And Equations**

**Statistical Analysis:**

After Data Were Collected, They Were Coded And Transferred Into Specially Designed Formats So As To Be Suitable For Computer Feeding.

Following Data Entry, Checking, And Verification Processes Were Carried Out To Avoid Errors During The Data Entry. Frequency Analysis, And Manual Revision Were All Used To Detect Any Errors.

The Statistical Package For Social Sciences (SPSS Version 21) Was Utilized For Both Data Presentation And Statistical Analysis Of The Results.

The Following Statistical Measures Were Used

- Descriptive Measures Included: Count, Percentage, Arithmetic Mean And Standard Deviation.
- Statistical Test Included: Pearson Correlation Coefficient For The Analysis Of Qualitative Variables.
- The Level Of Significance Selected For This Study Was P Less Than 0.05.

#### **V. Figures And Tables**

Table 1 Portrays The Characteristics Of Children With SCA. It Was Noticed That More Than Two Third Of Children's Age (68%) Ranged From 10-12 Years, While, 23% Of Them Their Ages Ranged From 8 To Less Than 10 Years And 9% Of Them Their Ages Ranged From 6 To Less Than 8 Years. It Was Also Shown That Nearly Three Quarters Of Children (73%) Were Male And 27% Of Them Were Female. Regarding To The Level Of Education, It Was Found That 79% Of Children Were In Primary Schools, 14% Of Them Were In Preparatory School, While Small Percent Of Children (7%) Didn't Join School. The Birth Order Of Children Was The Third And More For 47%, The First For 38% And The Second For 15%. Nearly Two Third Of Children Were In The Rural Area And 36% Of Them Were In The Urban.

Concerning To The Clinical Data As Reported By Studied Children, It Was Found That The Onset Of Illness Was One Year For 88% Of Children, While It Was Two Years For 12% Only Of Them. The Level Of Hemoglobin Was Low (From 8 To Less Than 11g/Dl) For The Majority Of Children (77%) And It Was Ranging From 11-13 G/Dl For 23% Of Them. It Was Shown That Most Of Children (92%) Received Blood Transfusion In 3 Months Intervals For 62% Of Children Or In 6 Months Intervals For 38% Of Them. Plus, Blood Transfusion Side Effects Were Found For 35.9% Of Children That Were In The Form Of Swelling And Pain At The Inserted Area Of Infusion (45.5%), Itching (33.3%) And Rash (21.2%). The Children's Dealing With Such Side Effects Was Either By Taking The Prescribed Medication ( 54.5%) Or Cold Compresses Application (45.5%). It Was Noticed Also That The Reported Received Treatment By Children Was Folic Acid (100%), Analgesic (100%), Antibiotic (74%), Iron (67%) And Vitamin B (62%). While, 5% Of Children Only Received Iron Chelation Therapy.

Characteristics Of Children's Mothers Are Presenting In Table 2, It Was Clear That More Than Half Of Mothers' Age(57%) Ranged From 40 Years And More, While, 31% Of Them Their Ages Ranged From 35 To Less Than 40 Years And 12 % Of Them Their Ages Ranged From 30 To Less Than 35 Years. Concerning Level Of Education, It Was Noticed That One Third Of Mothers (32%) Were Able To Read & Write, 25% Of Them Had Primary School And 16% Of Them Had Secondary School. While 13% Of Mothers Only Had High Education (University). It Was Seen That More Than Half Of Mothers (56%) Were Housewife And 44% Of Them Were Working.

Table3 Portrays The Percentage Distribution Of Children With Sickle Cell Anemia Regarding To Their Reported Dealing Approach Of Disease Symptoms. Unfortunately, It Was Found That Most Of Children Were Never (77%) Use Comfortable Measures In Dealing With Their Headache. While, The Majority Of Them Were Taking Either Drinks Sometimes (85%) Or Analgesic Often (81%).

Using Wet Cloth Were Done Sometimes By 92% Of Children Or Taking Medication Often By 100% Of Them For Treating Their Fever. Avoiding Tight Cloth Or Cold Application Was Done Sometimes By 86% Of Children In Dealing With Their Arms And Legs Swelling. While, 64% Of Them Were Often Following The Doctor's Orders.



Using Non Pharmacological Agents Such As Hot Application, Massage, Relaxation Technique Or Distraction Were Done Sometimes By 87% Of Children In Dealing With Their Bone And Joint Pain. While, 95% Of Them Were Often Taking Medication According To Doctor's Order, The Mean Score  $\pm$  SD 75.61 $\pm$ 15.19.

Table 4 Presents The Percentage Distribution Of Children With Sick Cell Anemia Regarding To Their Reported Experienced Pain. Unfortunately, It Was Shown That High Percent Of Children Were Never Experiencing Mild Pain (82%). While, The Experienced Pain Were Often Reported By Them As Moderate (89%) Or Severe (39%).

The Sites Of Feeling Pain That Were Often Reported By Studied Children Were Legs (80%), Whole Body (68%), Back (57%) And Arms (54%). While, Other Sites That Were Sometimes Reported By Them Were Stomach (59%) And Chest (53%), The Mean Score  $\pm$  SD 45.77 $\pm$ 6.60.

Figure1 Illustrates The Percentage Distribution Of Children With Sick Cell Anemia Regarding To Their Pain Intensity. It Was Shown That More Than Three Quarters Of Children (78%) Had Severe Pain And 22% Of Them Only Had Moderate Pain. While, None Of Them Had Mild Pain.

Table 5 Presents The Correlation Between The Mean Scores Of Children And Their Mothers' Reports In The Different Categories Of Quality Of Life. It Was Illustrated That There Were Positive Correlations And Significant Associations Between The Children Reports And Their Mothers' Reports In Different Categories Of Quality Of Life Except One (Intellectual Aspect). It Was Noticed That The Mean Scores Of Children's Reports Were More Than Their Mothers Reports In Most Categories; Dietary Habit[(58.80 $\pm$ 27.97), (52.14 $\pm$ 12.80), (R=0.677\*, P=<0.001\*)], Physical Activities[(27.67 $\pm$ 25.62), (21.25 $\pm$ 12.83), (R=0.494\*, P=<0.001\*)], Sleeping And Rest[(58.80 $\pm$ 27.97), (57.80 $\pm$ 26.31), (R=0.606\*, P=<0.001\*)], Social State [(19.58 $\pm$ 21.23), (18.25 $\pm$ 21.24) (R=0.442\*, P=<0.001\*)], Communication Skills[(55.75 $\pm$ 31.44), (46.08 $\pm$ 19.15), (R=0.641\*, P=<0.001\*)], Dealing With Symptoms Of The Disease[(75.61 $\pm$ 15.19), (63.61 $\pm$ 12.11), (R=0.487\*, P=<0.001\*)] And Experienced Pain [(45.77 $\pm$ 6.60), (42.09 $\pm$ 7.66), (R=0.298\*, P=<0.001\*)]. While, The Mean Scores Of Mothers' Reports Were More Than Their Children Reports Only In Two Categories; Emotional State [(24.58 $\pm$ 19.83), (22.71 $\pm$ 4.52), (R=0.407\*, P=<0.001\*)] And Treatment Process [(56.56 $\pm$ 19.08), (54.18 $\pm$ 7.61), (R=0.393\*, P=<0.001\*)].

It Was Found That The Level Of Quality Of Life For Studied Children Was Poor For More Than Two Third (66%) And Neutral For 23% Of Them. While, Small Percent Of Children (11%) Had Good Quality Of Life As Clarified In Figure 2.

It Was Illustrated From Table 6 That There Were Positive Correlations And Significant Associations Between The Children's Reports And Their Level Of Quality Of Life In Most Categories [Dietary Habit, Rest And Sleep, Intellectual Aspect, Social State, Communication Skills And Dealing Approach Of Disease Symptoms And Treatment Process [(R= 0.931\*, P= <0.001\*), (R=0.931\*, P=<0.001\*), (R=0.495\*, P=<0.001\*), (R= 0.263\*, P=0.008\*), (R= 0.972\*, P=<0.001\*), (R=0.963\*, P=<0.001\*), (R=0.295\*, P=0.003\*)] Respectively]. While, Correlations Were Not Found In Two Categories Only [Emotional Aspect And Pain Experienced] Where All Children Have Been Plotted In Poor Level (100%).

### III. Discussion

Sickle Cell Anemia Is Still A Significant Public Health Issue In Underdeveloped And Developing Countries. Worldwide, It Is Considered One Of The Most Common Inherited Diseases. It Is One Of The Diseases That Affects School-Age Children's QOL In Different Aspects. Consequently, The Goal Of Effective Management Of SCA Is To Allow Children To Function With Minimal Restrictions And Enjoy A Good Quality Of Life Throughout Their Lives<sup>(16)</sup>.

Quality Of Life Is A Broad Multidimensional Concept That Usually Includes Subjective Evaluations Of Both Positive And Negative Aspects Of Life <sup>(17)</sup>. In This Context, The Results Of The Present Study Revealed That Most Of Children With SCA Had Either Poor (66%) Or Neutral (23%) Quality Of Life. While, Only Small Percent Of Them Had Good Quality Of Life As Clarified In Figure 2. Panepinto Et Al. (2015)<sup>(18)</sup> Supported These Findings Where They Mentioned That Chronic Health Problem Such As SCA Is Negatively Affect The Health Related QOL Of Children. Other Authors Also Stated That The Majority Of Children Cases With SCA During School Years Had Low QOL<sup>(19)</sup>. On The Other Hand, Graves Et Al. (2016)<sup>(20)</sup> Clarified That Children With SCA Have Neutral QOL About Three Time More Than Children Who Suffer From Other Types Of Anemia.

The Children's Poor QOL That Was Shown In This Study Could Be Related To Multi-Factors Such As Dilatory Habits, Experienced Pain And Physical Activities. The Findings Of Current Study Showed Children's Poor QOL Regarding To Their Dilatory Habits 95.5% (Table 6), As Children Were Taking The Diet That Not Containing A Balanced Amount Of Iron, Folic Acid, Vitamin C, Copper And Cobalt. It Is Stated That These Elements Are Essential For Rbcs Formation And Their Deficiencies Will Lead To Distraction And Anemia. Minerals Such As Iron Is Important For Formation Of Hem Part Of Hemoglobin, Vitamins B12 And Folic Acid



Are Especially Essential For Final Maturation Of Rbcs, Copper And Cobalt Act As Catalysts In Hemoglobin Syntheses<sup>(21)</sup>. The Findings Of Present Study Were In Agreement With Lothe Et Al. (2015)<sup>(22)</sup> Who Revealed That Most Of Children With SCA Had Poor Nutrition And Reduction In Dietary Intake Of Iron, Vitamin A,C And B6 As Well As Protein And Iodine. While, Samira Et Al. (2015)<sup>(23)</sup> Reported That About One Third Of Children Had Good Dietary Habit But They Required Special Diet And Nutritional Supplements Daily As Folic Acid And Zinc.

The Experienced Pain The Children Had, As Illustrated In This Study Showed That 82% Of Children Were Never Had Mild Pain And 68% Of Them Were Often Feeling Pain On Their Whole Body (Table 4). These Results Were Congruent With Hamed (2016)<sup>(24)</sup> Who Found That Children With SCA Had Severe And Moderate Pain (46% And 54% Respectively). Montalembert Et Al. (2010)<sup>(25)</sup> And Laroche Et Al. (2012)<sup>(26)</sup> Stated That All Children Had Pain And Swelling In Bone Of Legs And Arms Indicated A Blockage Of Blood Vessel. Based On Literature, Sickle Cell Pain Is Worse Where Its Intensity Or Severity Episodes Vary Widely In Children. It Is Involved The Bone Pain As In Humerus, Femur, And Vertebrae That Is Presenting With Acute Tenderness, Swelling, And Fever. These Will Effect The Children's QOL Whereas It Hinders Their Social Interaction, Communication Skills And Physical Activity<sup>(27)</sup>

Moreover, Children With SCA Are Frequently Complain Of Difficulty In Sleep Initiation, Poor Sleep Quality And Excessive Daytime Sleepiness. The Present Study Agreed With Such Issues Where It Was Shown That Majority Of Children Had Poor QOL Regarding To Their Sleeping And Rest 95.5% As Shown In Table 6. In This Aspect, Alharbi Et Al. (2016)<sup>(28)</sup> Founded That More Than Half Of Children With SCA (55%) Had Always Trouble In Sleeping Pattern. On The Other Hand, Barakat (2018)<sup>(29)</sup> Showed That 56% Of Children With SCA Were Sleeping Good At Night

Furthermore, This Study Showed Poor QOL Concerning The Children's Physical Activities 87.9% As Portrayed In Table 6. These Were Parallel With Elsayed And El-Gawad (2015)<sup>(30)</sup> Who Found That Most Of Children Were Always Felt Tired To Take A Shower, Do Chores Around The House And Sports .Similar Findings Were Observed Also By Asnani Et Al. (2009)<sup>(31)</sup> Who Mentioned That Children With SCA Scored Less In Physical Activity And Daily Functioning Independently. The Findings Of This Study Could Be Explained In The Light Of Physical Activities Were Influenced By Biological And Physiological Status Of Children With SCA Under Deoxygenation, Whereas Hemoglobin S May Polymerize Leading To Rbcs Hemolysis, Vaso-Occlusion And Metabolic Changes Occurring During A Physical Exercise May Initiate Frequently Complain Of Difficulty In Pain<sup>(32)</sup>IN ADDITION, MORE THAN HALF OF MOTHERS WERE EITHER READ AND WRITE (32%) OR HAD PRIMARY SCHOOL (25%) AS WELL AS 56% OF THEM WERE HOUSEWIFE AS SHOWN IN TABLE 2 OF THIS STUDY. THE POOR MOTHERS' EDUCATION AND KNOWLEDGE MIGHT CONSTRAIN THEM TO PROVIDE IMPROPER MANAGEMENT AND HINDER THEIR DEALING EFFICIENTLY WITH THE DISEASE PROCESS FOR THEIR CHILDREN. KEEPING MOTHERS ALSO AS HOUSEWIFES COULD LIMIT THEIR AWARENESS TOWARD CHILDREN'S CARE DEMANDS AS IT DOES NOT GIVE THEM THE ENOUGH OPPORTUNITIES TO COMMUNICATE WITH OTHERS AND IMPROVE THEIR SKILLS.

Therapeutic Management For Children With SCA Can Be Classified Into Two Groups, Pharmacological And Non-Pharmacological<sup>(9)</sup>. It Was Shown In The Present Study That Children Who Had Good QOL In Dealing With Their Main Symptoms Of The Disease Were 100% As Clarified In Table 6. These Could Be Related To Most Of Studied Children Were Sometimes Taking Some Drinks As Ginger, Mint, Lemon (85%) And 100% Of Them Were Often Taking Their Medication To Relief Headache. It Is Stated That Herbs Is Naturally Soothing Substance, It Can Alleviate The Inflammation And Temperature Rise That Is Often With Headaches<sup>(7)</sup>. Ameh (2014)<sup>(33)</sup> Also Found That Children With SCA Are Sometimes Taking Herbs To Relieve Pain. Furthermore, The Using Of Non-Pharmacological Agents Such As Massage, Relaxation Technique And Distraction Were Sometimes Followed By 87% Of Studied Children To Treat Their Bone And Joint Pain(Table3). Authors Cited That Massage Help Children To Relieves Muscle Spasm, Improve Circulation And Provides Cutaneous Stimulation. Distraction Is Helpful Particularly For Children To Divert Their Attention To Something Else Such As Using Singing Songs, Telling Stories, Watching TV Or Listening To Holy Quran. Relaxation Technique The Children Used Assisting Them To Be More Comfortable And Reduce Their Pain Sensation<sup>(34)</sup>. The Used Non Pharmacological Agent Were Consistent With Shafer Et Al. (2014)<sup>(35)</sup> Who Found That Children Used A Proper Approach In Dealing With Their Symptoms Such As Massage, Relaxation And Distraction. Hurtig Et Al. (2009)<sup>(36)</sup> Who Stated Also That High Percent Of Children Were Using Wet Cloth (Tipped Compresses) When They Have Fever.

The Current Study Showed That There Is A Positive Correlations And Significant Associations Between The Mean Scores Of Children And Their Mothers' Reports In Most Of QOL Categories. This Agreed With Silva (2012)<sup>(37)</sup> Who Reported That Parents Of Children With SCA Were Highly Correlated With Their Children In Social State, Emotional State And School Achievement While Lower Correlation Was Found In Physical Activities. Patel Et Al. (2016)<sup>(38)</sup> Also Found That There Were Moderate To Good Correlations



Between Children And Their Parents' Reports. The Shown Positive Correlations Between Children And Their Mothers' Reports Could Be Also Explained In The Light Of The School Age Children Are Competent In Describing And Reporting Their Disease Process. This Is Congruent With Piaget Theory, Where It Is Stated That School Age Children Are In Concrete Operational Stage Of Their Cognitive Development. They Are Able To Think And Give Reasons As They Develop Concept Of Cause Effect Relationship And Can Manipulate Symbols. They Can Gain New Knowledge And Develop More Efficient Problem- Solving Ability And Greater Flexibility Of Thinking<sup>(30)</sup>. This Is Documented Also By The Results Of This Study Where The Age Of 68% Of Children Was Ranging Between 10-12 Years (Table 1).

#### IV. Conclusion

Based On The Findings Of The Current Study, It Is Concluded That The SCA Is Greatly Affecting The Children's Quality Of Life. Whereas, It Was Found That More Than Two Third Of Studied Children Had Poor Quality Of Life And Neutral For Less Than Quarter Of Them. While, Small Percentage Of The Studied Children Had Good Quality Of Life.

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**Table 1: Characteristics Of Children With SCA And Their Clinical Data; N=100.**

Characteristics of Children and Clinical Data Items	% No (100)	%
<b>A. Biosociodemographic Characteristics</b>		
• Age:- From 6 year	9	9
8-year	23	23
10-12 year	68	68
• Sex:- Male	73	73
Female	27	27
• Level of Education:- Primary school	79	79
Preparatory school	14	14
Not join school	7	7
• Birth order:- First	38	38
Second	15	15
Third and more	47	47
• Residence:- Rural	64	64
Urban	36	36
<b>B. Clinical Data</b>		
• The onset of the illness; n=100 First year	88	88
Second year	12	12
Hemoglobin level :- 8 - 10 g/ dl	77	77
11- 13 g/ dl	23	23



• Blood transfusion:		
- Received	92	92
- Not received	8	8
• Interval of received blood transfusion pre year; n=92		
- 3 month	57	62
- 6 month	35	38
• Blood transfusion side effects:		
- Found	33	35.9
- Not found	59	64.1
• Types of side effects; n=33		
- Rash	7	21.2
- Itching	11	33.3
- Swelling and pain at inserted area of infusion	15	45.5
• Children's dealing with the side effects:		
- Take the prescribed medication	18	54.5
- Tipped compresses application	15	45.5
• The received treatment for sickle cell anemia; n=100		
- Folic acid	100	100
- Analgesic	100	100
- Antibiotic	74	74
- Iron	67	67
- Vitamin B	6	62

Table 2: Characteristics Of Children's Mothers; N=100.

Items	%
• Age of Mother:-	
From 30 years	12
35- years	31
40 years and more	57
• Level of Education:-	
- Illiterate	7
- Read and Write	32
- Primary school	25
- Preparatory school	7
- Secondary school	16
- Diploma	0
-University	13
• Mothers' work:-	
- Housewife	56
- Working	44

Table 3: Percentage Distribution Of Children With Sickle Cell Anemia Regarding To Their Reporting Dealing Approach Of Disease Symptoms; N=100.

Items	Never %	Sometimes %	Often %
•Headache:			
1.Using Comfortable Measures (E.G. Stay In Calm Place, Keep Away From Noise)	77	0.0	22
2.Taking Some Drinks (E.G. Ginger, Mint, Lemon)	4	85	11
3.Taking Analgesic	0.0	19	81
•Fever:			
1.Using Wet Cloth (Tipped Compresses)	0.0	92	8
2. Taking The Ordered Medication	0.0	0.0	100
•Swelling In The Arms And Legs:			
1.Avoiding Tight Clothing And/Or Cold Application	14	86	0.0
2.Following Doctor's Orders	21	15	64
•Pain In Bone And Joints:			
1. Using Non Pharmacological Agents (E.G. Hot Application, Massage, Relaxation Technique And Distraction)	13	87	0.0
2.Taking Medication According To Doctor's Order	0.0	5	95
Mean Scores± SD	75.61±15.19		

Table 4: Percentage Distribution Of Children With Sickle Cell Anemia Regarding To Their Reported



Experienced Pain; N=100.

Experienced Pain	Never %	Sometimes %	Often %
(A): Pain intensity:			
1. Mild	82	18	0.0
2. Moderate	0.0	11	89
3. Severe	29	32	39
(B): Sites of feeling pain:			
1. Whole body	0.0	32	68
2. Arm	3	43	54
3. Leg	0.0	20	80
4. Chest	29	53	18
5. Back	0.0	43	57
6. Stomach	25	59	16
Mean score $\pm$ SD	45.77 $\pm$ 6.60		

Figure (1): Percentage Distribution Of Children With Sickle Cell Anemia Regarding To Their Reported Pain Intensity

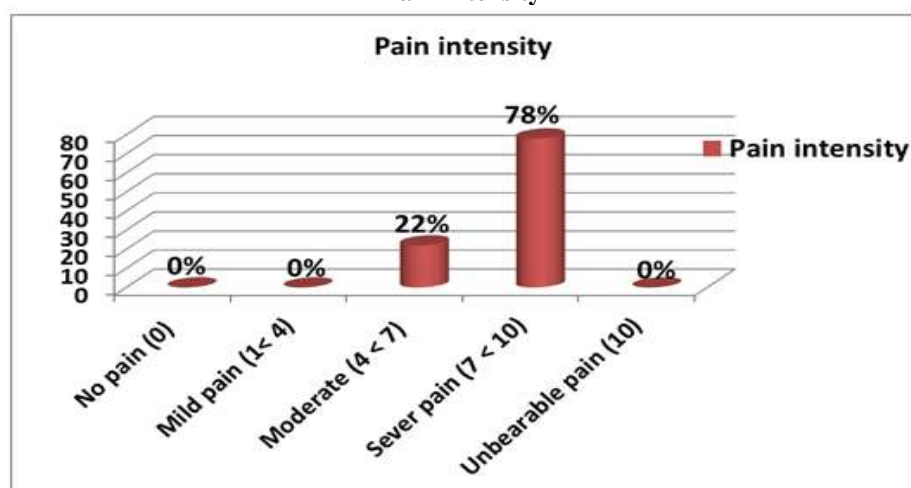
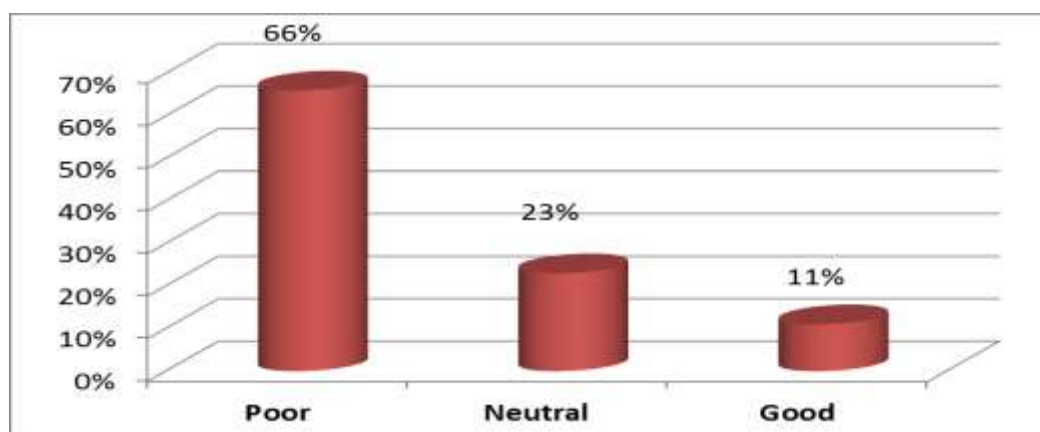


Table (5): Correlation Between The Mean Scores Of Children And Their Mothers' Reports In The Different Categories Of Quality Of Life

Items	Mean % Score		R	P
	Children's	Children's Mother		
Dietary Habit	58.80 $\pm$ 27.97	52.14 $\pm$ 12.80	0.677*	<0.001*
Physical Activities	27.67 $\pm$ 25.62	21.25 $\pm$ 12.83	0.494*	<0.001*
Rest And Sleep	58.80 $\pm$ 27.97	57.80 $\pm$ 26.31	0.606*	<0.001*
Intellectual Aspect (N = 93)	28.06 $\pm$ 27.04	19.46 $\pm$ 20.07	0.082	0.434
Emotional State	22.71 $\pm$ 4.52	24.58 $\pm$ 19.83	0.407*	<0.001*
Social State	19.58 $\pm$ 21.23	18.25 $\pm$ 21.24	0.442*	<0.001*
Communication	55.75 $\pm$ 31.44	46.08 $\pm$ 19.15	0.641*	<0.001*
Dealing Approach Of Disease Symptoms	75.61 $\pm$ 15.19	63.61 $\pm$ 12.11	0.487*	<0.001*
Pain Experienced	45.77 $\pm$ 6.60	42.09 $\pm$ 7.66	0.298*	0.003*
Treatment Process	54.18 $\pm$ 7.61	56.56 $\pm$ 19.08	0.393*	<0.001*

Figurer (2): The Level Of Quality Of Life For Children With Sickle Cell Anemia





**Table (6): Correlation Between The Children Report And Their Level Of Quality Of Life In Deferent Categories.**

Items Of Quality Of Life	Level Of Quality Of Life						R	P
	Poor (N = 66)		Neutral (N = 23)		Good (N = 11)			
	N	%	N	%	N	%		
Dietary Habit	63	95.5	1	1.5	2	3.0	0.931*	<0.001*
Physical Activities	58	87.9	1	4.3	2	18.2	0.151	0.133
Sleep And Rest	63	95.5	0	0.0	11	100	0.931*	<0.001*
Intellectual Aspect (N = 93)	62	93.9	4	19.0	5	83.3	0.495*	<0.001*
Emotional State	66	100	0	0.0	0	0.0	-	-
Social State	66	100	0	0.0	2	18.2	0.263*	0.008*
Communication	65	98.5	0	0.0	11	100	0.972*	<0.001*
Dealing Approach Of Disease Symptoms	2	3.0	0	0.0	11	100	0.963*	<0.001*
Pain Experienced	66	100	0	0.0	0	0.0	-	-
Treatment Process	55	83.3	7	30.4	0	0.0	0.295*	0.003*

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