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Clinical Assessment of Structural Changes in Vankshan Sandhi (Hip Joint) with Respect to Avascular Necrosis in Sickle Cell Anemia: A Prospective Observational Study

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Vd. Priya Ashokrao Thakare*[1], Sudam. L. Kate[2], Girish T. Kulkarni[3], M.N. Suryawanshi[4], Shubhangi Jadhav[5]

1 Research Officer (Ay.), Regional Ayurveda Research Institute, Nagpur, Maharashtra, India. 2 HOD Sickle Cell Department, MAM's Sumatibhai Shah Ayurved Mahavidyalaya, Malwadi, Hadapsar, Pune, Maharashtra, India. 3 Assistant Professor Anatomy, MAM's Sumatibhai Shah Ayurved Mahavidyalaya, Malwadi, Hadapsar, Pune, Maharashtra, India. 4 Assistant Director Incharge, Regional Ayurveda Research Institute, Nagpur, Maharashtra, India. 5 Assistant Professor, Dravyaguna Department, Bhausaheb Mulak Ayurved College& Research Hospital Buttibori, Nagpur, Maharashtra, India.

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ABSTRACT

Background: Sckle Cell Anaemia is a hereditary disorder and is very common amongst Scheduled Tribes (ST) and Scheduled Caste (SC) of Satpuda hilly ranges of North Maharashtra. The prevalence rate is 22.1% carrier (A+S pattern) and 1.25% sufferer (S+S pattern). Material and Methods: Total 30 patients of SCA were selected of age group between 15-40 yrs on the basis of selection criteria. Goniometry is used to study structural changes and movements at hip joint. Clinical assessment criteria in Sickle Cell Anaemia was assessed. Results: Leg length measured and compared with each other. Difference between leg length of right and left leg was statistically not found significant. Range of movement of each movement at hip joint was measured with the help of universal goiometer. Observed range was compared with normal range and it proved significant. Conclusion: Total 30 patient of age group 15-40 of both the gender were selected for study. Measuring (degree of) hip joint movement with Goniometer and other parameters like pain, oedema, local temperature, difficulty in squating, lurching gait, leg length are used in this study on Anukta Vyadhi (Sickle Cell Anameia) causing Kshayatmak Vikruti of Dhatu. Consequences of this Anukta Vyadhi are observed in Asthi and Sandhi. Structural changes in Vankshan Sandhi in Sickle cell patients were observed with help of radiology. Similarly clinical assessment criteria for structural changes in Vankshan Sandhi were observed. We found that due to lack of advanced technology at these remote locations and for patients with poor economy this clinical assessment criterion will be of gold standardto diagnose such a painful life threatening condition (AVN).

INTRODUCTION:

Sickle Cell Anaemia is red blood cell single gene incurable disease. SCA is a result of a mutation in the β chain of haemoglobin molecule where 6th amino acid - Glutamic acid is replaced by - Valine. Red cells with this type of haemoglobin become sickle shaped when deoxygenated, causing changes in blood viscosity and red cell membrane. Alteration in the shape and flexibility of RBC's affect their ability to deliver oxygen to tissues which creates many complications. There is no permanent solution to stop the sickling of cells or to provide them their original form.

History Of The Disease:

In 1910, Dr. James B. Herrick of Chicago America noticed sickle shaped red blood corpuscles while examining the blood of a patient of severe anaemia. Clinical findings of which were later labeled as 'Sickle Cell Anaemia'. Mason introduced the term 'Sickle Cell Anaemia' in 1922. It was shown in 1923 by Huck that the sickling phenomenon was unquestionably inherited. Hahn and Gillespe in 1927 showed that sickling developed as a result of a fall in the partial pressure of O2. In 1949, Pauling Etal demonstrated the sickling phenomenon is associated with abnormal haemoglobin.

Epidemiology -

Hemoglobinopathies are especially common in areas where malaria particularly falciparum trait is endemic. The patient with sickle cell trait is relatively resistant to the lethal effects of falciparum malaria in early childhood. Sickle cell disease is the most common structural hemoglobinopathy occurring in heterozygous form in about 20% in topical Africa & 8% of American blacks and in homozygous form in 1 in 400. Sickle cell disease is not limited upto a particular society, religion or country. It has been seen in South and North America, Cuba, Jamaica, Portugal, Cyprus, Italy, Turkey, Israel, **India**, South Arabia and Shrilanka. In **India**, Rajasthan, Gujarat, Maharashtra, Madhya Pradesh, Chhattisgarh, Orissa in high percentage while Andhra Pradesh, Jharkhand, Uttar Pradesh states and there border areas are also affected. In India Sickle cell disease is found specifically in SC, ST and Other backward communities.

Prevalence-

The inherited disorders of hemoglobin are the commonest monogenic disorders in India. Of these, a-thalassemia is seen practically in every community of the country with varying frequency while sickle gene is confined mainly to dravidians and predravidians tribes inhabiting malaria endemic regions. Duringthe last 54 years, several groups of investigators conducted hospital-based or epidemiological surveys in various ethnic groups. Based on these surveys, prevalence of sickle gene is found to be 0-18% in northeastern India, 0-33.5% in western India,22.5-44.4% in central India and 1-40% in southern India and the gene frequency of Hb-S varies between 0.031- 0.41. Sickle gene was first detected by Lehman and Cutbush (1952) among the tribals of Nilgiri Hills. Since then, more than 300 tribal groups have been screened to look for the presence of sickle gene (Bhatia & Rao 1987, Balgir 1996, Mohanty & Mukherjee 2002).The prevalence varies considerably among different tribal groups ranging from 0-35%.

The magnitude of the problem-

About 7% of the world's population is carriers of some form of hemoglobin disorder. There are about 270 million carriers of sickle cell anemia and/or thalassemia. (WHO1994). In India, sickle cell gene is mainly restricted to tribal and scheduled caste population where carrier frequencies range between 5-40% or more with three focal points (Bhatia and Rao 1987). Kate (2000) compiled the data generated by various groups through screening various tribal populations from Maharashtra. This revealed that average prevalence ofsickle cell carrier among the tribal population was 10% and that of homozygotes was0.5%. Considering the tribal population of the state as 90 lakhs (census 1991), the expected carriers of sickle cell would be 9 lakhs and expected number of homozygotes would be 45000. Sharma et al (1986) in 1812 cases of SCD have reported that there were 33 deaths (1.81%) due to SCD. The causes of mortality were septicemia, acute splenic sequestration, severe anemia and hemolytic crisis. Improvement in the life style of the tribals would help to control morbidity and mortality especially among sicklers.

Global burden of disease - In 2006, the World Health Organization (WHO) recognized SCA as a global public health problem[3]. In 2010, the 63rd World Health Assembly adopted a resolution on the prevention and management of birth defects, including sickle cell disease and thalassemias. Finally, hemoglobinopathies have been included in the most recent Global Burden of Diseases, Injuries, and Risk Factors Study (the GBD 2010 study), which aims at

providing a comprehensive and systematic evidence-based assessment of the burden of major diseases and injuries[4].

It is anticipated that as overall under 5 mortality U5M begins to fall because of improved nutrition and medical facilities, an increasing proportion of children under 5 yrs with SCA will survive long enough to reach medical attention. Public health improvements (including the widespread use of prophylactic penicillin and vaccination) will help an increasing proportion of these children to survive through childhood and adulthood, and therefore to present for diagnosis and treatment (e.g., hydroxyurea, hospitalization, transfusion) generating lifelong costs. The lack of interventions will inevitably lead to a large burden on the public health infrastructures and budgets of the countries most affected[5,6].

Skeletal Changes in SCA -

Sickle Cell Anaemia is a multisystem disorder in which skeletal changes are observed. The 1st report of a pathologic study of bone changes in SCA was by George S. Graham in 1924[7]. Skeletal changes in Sickle Cell Haemoglobinopathies occur mainly because of hyperplasia of bone marrow and vascular insufficiency resulting in thrombosis and infarction. Associated bone infection is quite common leading to Osteomyelitis and sequestration. Because of hyperplasia of erythrocytes, there is an increase in blood viscosity, stasis capillary thrombosis and finally infarction of bone.

The initial infarcts occur in the most distal portion of bones (i.e. femoral head), in the subchondral area, where there is maximum sickling and where the circulation through the collaterals is very poor and limited. The blood supply is reduced to the cartilages and bone and it shows the dead bone which is avascular and impaired. Changes are seen in – Long bones, Skull, Vertebrae, Short bones of the hand and feet, Ribs and Scapulae. Changes due to vascular insufficiency like Avascular Necrosis are seen in humeral head also. Roentgen findings in skull includes – Thickening of the outer table, Hair on end appearance and marked osteoporosis. In long bones there is marked thinning and irregularities of Cortex, prominent trabeculations, expansion of medullary cavity.

Skeletal changes in sickle cell hemoglobinopathy develop as a result of two pathological processes : 1) Chronic haemolytic anaemia, secondary to spontaneous erythrocyte sickling and premature cell death leading to bone marrow hyperplasia. 2) Sickling resulting in tissue ischemia and bone infarction secondary to small vessel thrombosis. Osteonecrosis of the

femoral head is very common presentation of SCD. The high incidence of the femoral head is very common presentation of SCD. The high incidence can be explained on the peculiar vascular supply of femoral head where single vessel- the Ascending branch of lateral femoral circumflex artery or one of its branches if affected can lead to Osteonecrosis of the femoral head.

Avascular Necrosis

Osteonecrosis also referred to as avascular necrosis (AVN), aseptic necrosis, and ischemic necrosis is not a specific disease but rather a condition in which a circumscribed area of bone becomes necrotic as a result of a loss of its blood supply. Avascular necrosis is a disease where there is cellular death (necrosis) of bone components due to interruption of the blood supply. Without blood, the bone tissue dies and the bone collapses. If avascular necrosis involves the bones of a joint, it often leads to the destruction of the joint articular surfaces.

There are two forms of osteonecrosis.

1) Traumatic 2) Atraumatic

The traumatic form has a definitive causal event and is isolated to the particular injured bone. The atraumatic form has multiple etiologies and can involve multiple bones.

Epidemiology of AVN

It is currently estimated that 15,000 to 20,000 new cases are diagnosed annually in the United States alone and that osteonecrosis accounts for approximately 10% of the total hip replacements (THRs) performed.

Etiology of AVN - Many etiological factors can be identified in patients with nontraumatic osteonecrosis.

These include

1) Intra-osseous marrow displacement disorders- that lead to increased pressure in the femoral head and neck. Such as- Gaucher’s disease, leukemia, and myeloproliferative disorders.

2) Conditions that lead to direct cell toxicity- Radiation, Chemotherapy.

3) Mechanical blockage of vessels may be caused by - emboli composed of abnormal red blood cells such as in- Sickle cell disease and thalassaemia; or - Nitrogen bubble emboli- such as in caisson disease (decompression sickness) or dysbarism.

4) Risk factors also include-

- Alcoholism
- Excessive steroid use
- Vascular compression
- Hypertension.

5) In some cases it is idiopathic.

Presentation of disease-

While it can affect any bone, about half of cases show multiple sites of damage, avascular necrosis primarily affects the joints at the shoulder, knee, and hip. The classical sites are: **head of femur, neck of talus and waist of the scaphoid**.

Clinical avascular necrosis most commonly affects the ends (epiphysis) of long bones such as the femur. Other common sites include the humerus , knees, shoulders, ankles and the jaw.

SCA from Ayurvedic Perspective-

Sickle Cell Anamia the term is not mentioned In Ayurvedic text so to study this disease doshadushti is to be studied extensively. Similarly its inheritance is also to be studied from Ayurveda perspective-

Genetics in Ayurveda

In Ayurveda very few diseases are explained as genetic or Sahaj Vyadhior Adibalapravrutta Vyadhi[7]. Adibalaravruttivyadhi is described to any inherited defect in the *Shukra* and/ or *Shonit*(both of which collectively form the new living beings). If Vatadidosha present in the Shukra (and/or Shonit) get vitiated, then accordingly they produce diseases in the upcoming generations. Apart from diseases mentioned under this category in Samhitas- some other disorders also seems to be genetic as per modern considerations, like Sickle Cell Anaemia.

This study is focused on structural changes occurring at Vankshan Sandhi i.e. Hip joint in SCA patients which is important aspect of the locomotor system.

Place of Study- MAM's Sickle Cell Unit, Dhadgaon, Nandurbar district.

MAM i.e. Maharashtra Arogya Mandal, Hadapsar, Pune, is a Non Governmental organization (NGO) established in June1960 by late **Dr. S.T. alias Dadasaheb Gujar** and his colleagues with the aim to provide better health care facilities to the poor and underprivileged classes of society. Sickle Cell Anaemia Project is run by MAM since 1998 with a mandate to provide **diagnosis, treatment, counseling and prevention.** This Institute is working in North Maharashtra (Satpuda region) for last 23 years and found prevalence to be Carrier 22% and Sufferer 1.5%.

All complications recorded in the medical literature were observed in our patients, except in Leg Ulcer. A single gene defect affect most of the important ogans of the body. Sickle Cell Crisis is hallmark of the disease it give rise to tremendous unbearable pain. We have establish a community control programme Centre (Sickle Cell unit) under supervision of Dr. S. L. Kate in remote tribal area i.e. Village Roshmal BK, Taluka Dhadgaon,Dist. Nandurbar Maharashtra (21.8263N and 74.2172 E). This centre provides following facilities-

Diagnosis

Clinical Examination and Free Medication

Counseling

Preventive guidelines

We develop simple laboratory technology i.e. Solubility test followed by Electrophoresis for diagnosis. We also have developed Ayurvedic drug known as SC3 containing six polyherbal medicine. The drug trial has been taken by AYUSH Dept. of Govt. of India for three years and approved this drug for sickle cell patients. We have more than 5000 sickle cell patients using this drug and are satisfied about results.

Osteonecrosis is common among Sickle Cell patients. We observed Avascular necrosis femoral head (AVNFH), Avascular necrosis humeral head (AVNHH),and Fish Mouth Vertebra. Among these AVNFH is most common. About 10% of patients visiting our centre

suffer from AVNFH pain and since there is no medical facilities available in remote hilly area, patient remains affected throughout the life.

The said project is completed at Dhadgaon. It was an observational study. Pathophysiology of Sickle Cell Anaemia is being extensively studied in modern science. Systematic treatment along with bone marrow transplant is also available and further genetic studies are still going on as no permanent cure is available yet. Main reason behind the selection of this topic for research is Sickle Cell Anaemia is yet to be studied so far in Ayurveda and to introduce some treatment for any disease one has to study its Samprapti (etiopathology). This disease is not mentioned by name in Ayurvedic literature so it has to be studied as AnuktaVyadhi. So a humble effort is put forth.

MATERIAL AND METHODS

Data Collection-

- Literary review- From Ayurvedic and Modern textbooks and e literature
- CRF designed to record patients data

Study Design- Observational Study

Sampling- Simple Random Sampling

Ethical Clearance- Ethical clearance was obtained from Institute's Ethical Committee before the initiation of trial.

Selection Criteria

Inclusion Criteria –

- Age - 15 – 40 yrs
- Sex – Both Genders.
- Diagnosed patients of sickle cell anaemia suspecting avascular femur head necrosis.
- Guardian consent was taken wherever necessary.

Exclusion Criteria –

- Patients of femoral head necrosis other than Sickle cell anaemia.

- All anaemia patients of other than sickle cell anaemia
- Thalassemia.
- Accidental injury, fracture, dislocation.
- Rheumatoid arthritis, Osteoarthritis.
- Any major hormonal disorder.
- Any congenital anomaly.
- Dropouts –

- Irregular followup.
- Unwilling patients.
- Surgically corrected hip joint.
- Death.

Methodology

- Case record form was designed to assess changes in *Vankshan Sandhi* in Sickle Cell Anaemia patients.
- Informed written consent was obtained from every individual.
- This study was conducted in sickle cell department of MAM's Hadapsar college – Dhadgaon Unit.
- Oxford pain chart was used for pain assessment.
- Goniometer was used for measurement of angle of movement of *Vankshan Sandhi.*
- Their leg length was measured with measuring tape.
- Radiographs were taken to observe structural changes.

Goniometry

The term goniometry is formed by Greek words Gonio means angle and Metron means measures (Norkin and White, 1994). The amount of movement available at a joint is called

range of motion (ROM). Therefore Goniometry refers to the measurement of angles, in particulars the measurement of angles created at human joints by the bones of the body.

Three Types Of Goniometry

1. Universal 2.Gravity-dependent Goniometry 3.Electro Goniometry

Universal - It is the commonly used to measure joint position and motion. The name universal implies the wide use of instrument for almost all the joints. It is made up of metal/ steel/ plastic.

The goniometer is a standard device for measuring angles and is relatively inexpensive.

Goniometers come in various sizes for use with different joints.

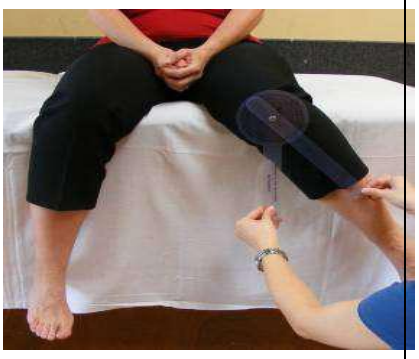
A goniometer has three components. The circular or semi-circular base with degree markings is called the body. Goniometers also have two arms. The stationary arm is fixed to the body. The moving arm is moved in concert with the patient's joint movement and points to the readings on the goniometer body.

Table No.1: Measuring Hip joint movement with the Goniometer

Sl. No.	**Movements of Hip joint**	**Image**
1	**Flexion** -Normal ROM: 110-120^0	
i.	Patient in supine position.	
ii.	Locate the greater trochanter of the femur and the lateral epicondyle of femur.	
iii.	Place the center of the goniometer body over the	

	greater trochanter. Align the stationary arm with the lateral mid-line of the pelvis. Align the moving arm with lateral epicondyle of femur.	
iv.	Stabilize pelvis and have patient flex hip bringing thigh close to the trunk with knee bent. Align moving arm with lateral epicondyle of femur. Read the angle.	
2	**Extension** Normal ROM: 10-15^0	
i.	Patient in prone position.	
ii.	Locate the greater trochanter of the femur and the lateral epicondyle of the femur.	
iii.	Place the center of the goniometer body over the greater trochanter. Align the stationary arm with the lateral mid-line of the pelvis.	
iv.	Stabilize pelvis and have patient extend hip by lifting the leg upward. Align moving arm with lateral epicondyle of femur. Read the angle.	
3	**Abduction** Normal ROM: 30-50^0	
i.	Patient in supine position, with toes pointing up (no rotation).	
ii.	Locate the anterior superior iliac spine (ASIS) on both sides and the midline of the femur.	
iii.	Place the center of the goniometer body over the ASIS of the side being measured and point the stationary arm to the opposite ASIS.	
iv.	Align the moving arm with the with the midline of the femur, using the midline of the patella for reference.	

v.	Stabilize pelvis and have patient abduct the leg, moving laterally outward. Read the angle.	
4	**Adduction** Normal ROM: 0-30^0	
i.	Patient in a supine position, with toes pointing up (no rotation). Have patient move the leg of the non-testing side laterally out of the way.	
ii.	Locate the anterior superior iliac spine (ASIS) on both sides and the midline of the femur, using the midline of the patella for reference.	
iii.	Place the center of goniometer body over the ASIS of the side being measured and point the stationary arm to the opposite ASIS.	
iv.	Align the moving arm with the with the midline of the femur, using the midline of the patellar for reference.	
v.	Stabilize pelvis and have patient adduct the leg, moving it medially inward. Read the angle.	
5&6	**Hip External and Internal Rotation**	
i.	Patient in seated position with lower legs dangling freely(short sit), perpendicular to floor.	
ii.	Locate the anterior aspect of the patella and the midline of the tibia.	
iii.	Place the center of goniometer body over the center of the patella and keep stationary arm of goniometer perpendicular to floor.	
iv.	For **external rotation** Stabilize distal femur head with one hand and distal fibular with another. Guide the patient to move the	

	lower leg medially as much as he/she can without moving the hip. Stationary arm is perpendicular to the floor: align the moving arm with the midline of tibia. Read angle. Normal ROM : 40-60^0	
v.	**For internal rotation**, Stabilize distal femur head with one hand and distal fibular with another. Guide the patient to move the lower leg laterally as much as he/she can without moving the hip. Stationary arm is perpendicular to the floor: align the moving arm with the midline of tibia. Read angle. Normal ROM : 30-40^0	

Assessment Criteria

1. Pain

Absent	No Pain	0
Present only on weight-bearing	Mild	1
Pain at rest also	Moderate	2
Restricted physical activities considerably or caused sleeplessness at night	Severe	3

2. Oedema

Absent	0
Slightly Present	1
Markedly Present	2
Severely Present	3

3. Local temperature

Normal	0
Mild	1
Moderate	2
Severe	3

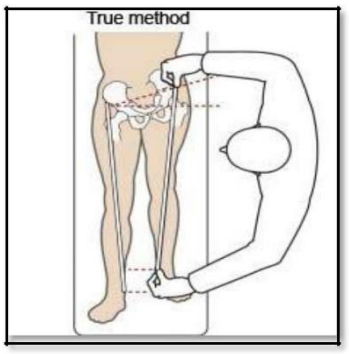
4. Degree of Hip-Joint movement:

Range of motion of hip joint measured with the help of goniometry.

5. Leg length:

Clinical Methods

The most commonly use methods for measuring leg length discrepancy are the apparent and true leg length measurements.



True leg length measurements

This method involves measuring from the protruding pelvis bone, anterior superior iliac (see red points on the diagram below) to the ankle joint, medial malleoli. The use of a tailor's tape measure is accepted.

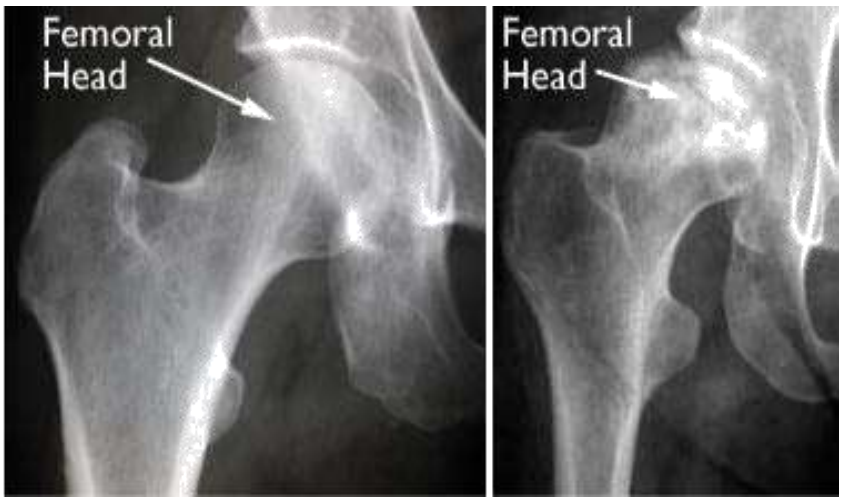
Position- Supine

By comparing the results for each leg the discrepancy can easily be found.

The apparent leg length measurement

This method is similar to the above, however, it usually gives slight less accurate results. To measure a leg length discrepancy using this technique, measure from the belly button (umbilicus) to the ankle joint, medial malleoli.

6. Radiological finding:



Normal XRay | SCA - XRay

Table No.2 FICAT classification for AVN Staging:

Stage	Clinical symptoms	X-ray
Stage 0	Nil	Normal
Stage I	Pain typically In the groin	Normal/ minor osteopenia
Stage II	Pain and stiffness	Mixed osteopenia &/ sclerosis&/subchondral cysts, without any subchondrallucency
Stage III	Pain and stiffness +/- radiation to the knee and limp	Crescent sign and external cortical prolapse
Stage IV	Pain and limp	End stage with evidence of secondary degenerative changes.

OBSERVATION

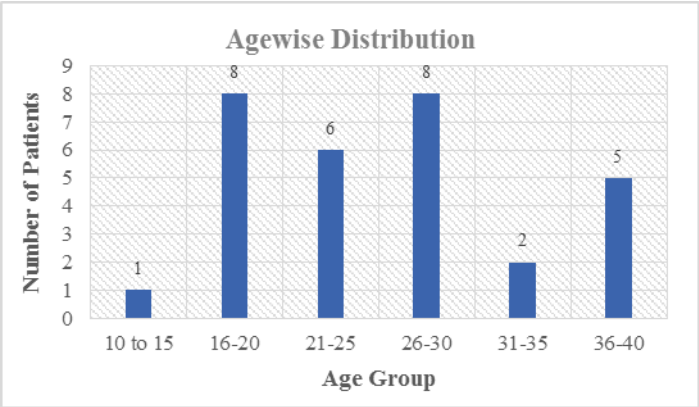
In the present study, 30 patients fulfilling inclusion criteria were selected & following results were incurred based on observations.

1. Age Wise Distribution Of Data

Incidence was observed to be highest in the age groups 16-20 and 26-30 constituting 8 patients (26.66%) in each group. Followed by 6 patients (20%) in 21-25 age group, followed by 5 patients (16.66%) in 36-40 age group.

Table no. 3 Frequency distribution table of Age wise distribution

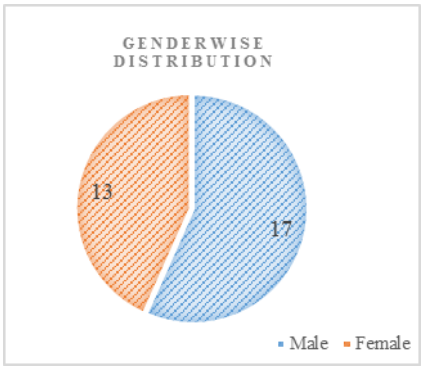
Age Group	No. Of Patients	%
10 to 15	1	3.33
16-20	8	26.66
21-25	6	20
26-30	8	26.66
31-35	2	6.66
36-40	5	16.66



Graph 1: Age wise Distribution of Data

2. Gender Wise Distribution Of Data

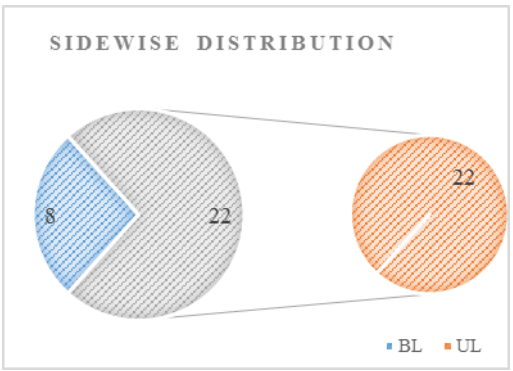
Data showed that there were 17 male patients (56.66%) while female patients were 13(43.33%).Hence it can be said that data is nearly equally distributed slightly more towards male patients.

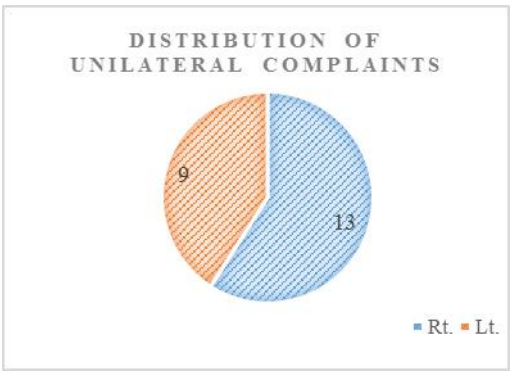


Graph 2: Gender wise Distribution of Data

3. Side Wise Distribution Of Data

Patients complaining of hip joint pain and other symptoms at hip were classified according to the affected side as if unilateral (UL) and bilateral complaints (BL). It was observed that 22 patients (73.33%) were complaining of unilateral involvement while 8 patients (26.66%) were bilaterally involved. Among unilateral involvement patients complaining for right side were 13(59.09%) more than that for left side 9(40.9%).





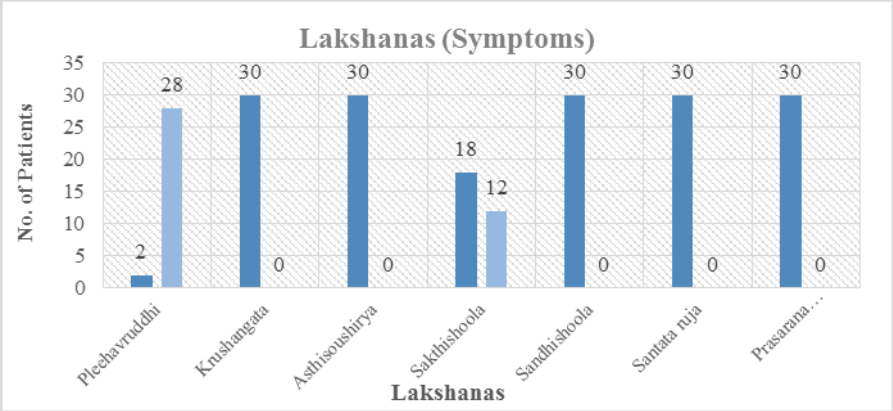
Graph 3: Sidewise Distribution of Data

4. Distribution of data based on symptoms.

Based on assessment criteria some symptoms were seen in all 30 patients (100%) of the study. These were- *krushangata, Asthisoushirya, Sandhishoola, Santataruja* and Prasarana Akunchana Pravruttisa Vedana. Pleehavruddhi was seen in only 2 patients (6.66%) and Sakthishool in 18 (60%).

Table 4 Frequency distribution Table Presenting Lakshnas (Symptoms) present.

Symptoms (Lakshanas)	Present		Absent	
	No. of patients	%	No. of patients	%
Pleehavruddhi	2	6.66%	28	93.33%
Krushangata	30	100%	0	0
Asthisoushirya	30	100%	0	0
Sakthishoola	18	60%	12	40%
Sandhishoola	30	100%	0	0
Santataruja	30	100%	0	0
Prasarana Akunchana Pravruttisa Vedana	30	100%	0	0



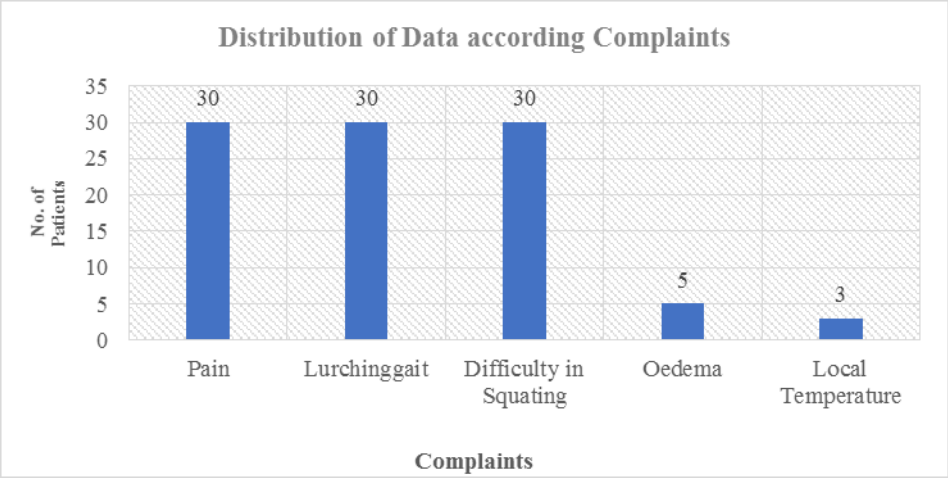
Graph 4: Distribution of Data based on Lakshanas present in them

5. Complaints

Data distribution shows that all 30 patients were having pain, lurching gait and difficulty in squatting. 5patients had oedema at *Vankshan sandhi* and local temperature was raised in 3 patients only.

Table 5 : Frequency Distribution Table Presenting Complaints

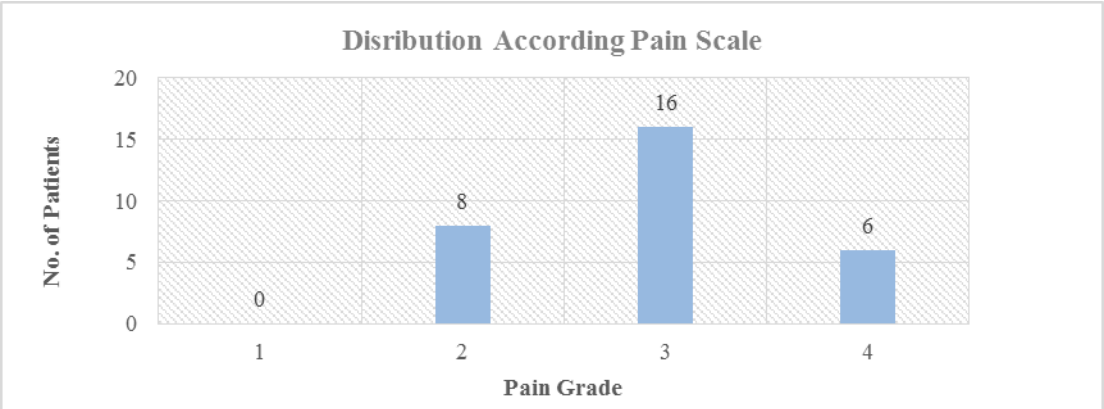
Complaints	No. of Patients	Percentage
Pain	30	100 %
Lurchinggait	30	100%
Difficulty in Squating	30	100%
Oedema	5	16.66%
Local Temperature	3	10%



Graph 5: Distribution of Data on the basis of Complaints

6. Distribution Of Data According Pain

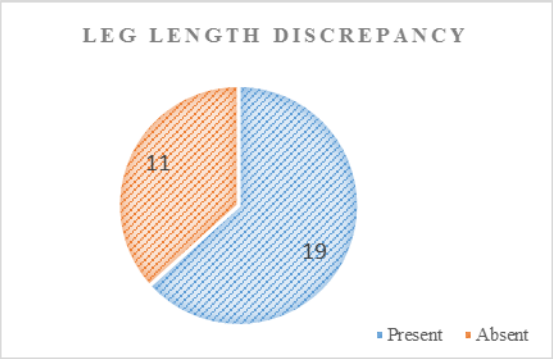
Pain scale observed as per the oxford pain chart. It was observed that 6 patients (20%) experience severe pain, 16 patients (53.33%) experienced moderate pain while 8 patients (26.66%) experienced mild pain.



Graph 6 – Graph of Distribution According to Pain Scale

7. Leg Length Discripencywise Distribution Of Patients

Leg length discrepancy was observed in 19 patients (63%) while 11 patients don't show any discrepancy.

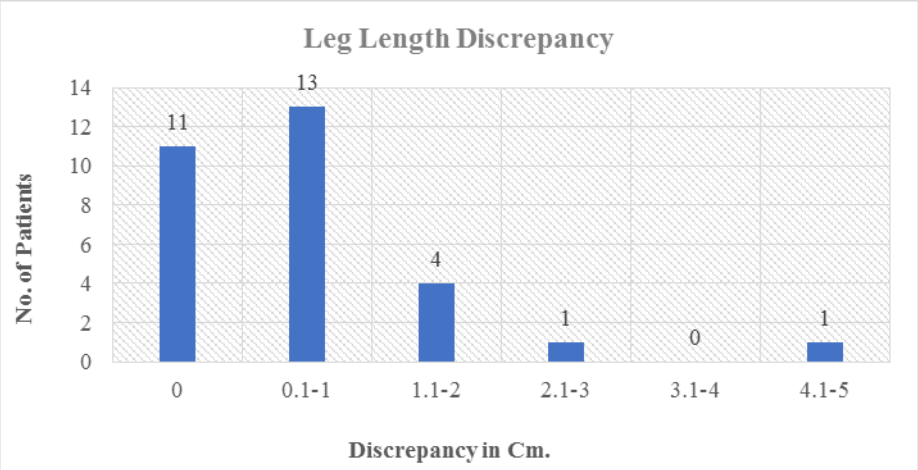


Graph 7 Leg Length Discrepancy

Out of 19 showing discrepancy most of the patients i.e. 13 undergo leg length shortening between 0.1 to 1 cm; 4 patients undergo 1.1-2cm difference; while 1 patient had 3cm and another one 5cm difference.

Table 6- Frequency Distribution Table of data as per Leg length Discrepancy in cm.

Class (Discrepancy in Cm.	Number of Patients	%
0	11	37
0.1-1	13	43.33
1.1-2	4	13.33
2.1-3	1	3.33
3.1-4	0	0
4.1-5	1	3.33



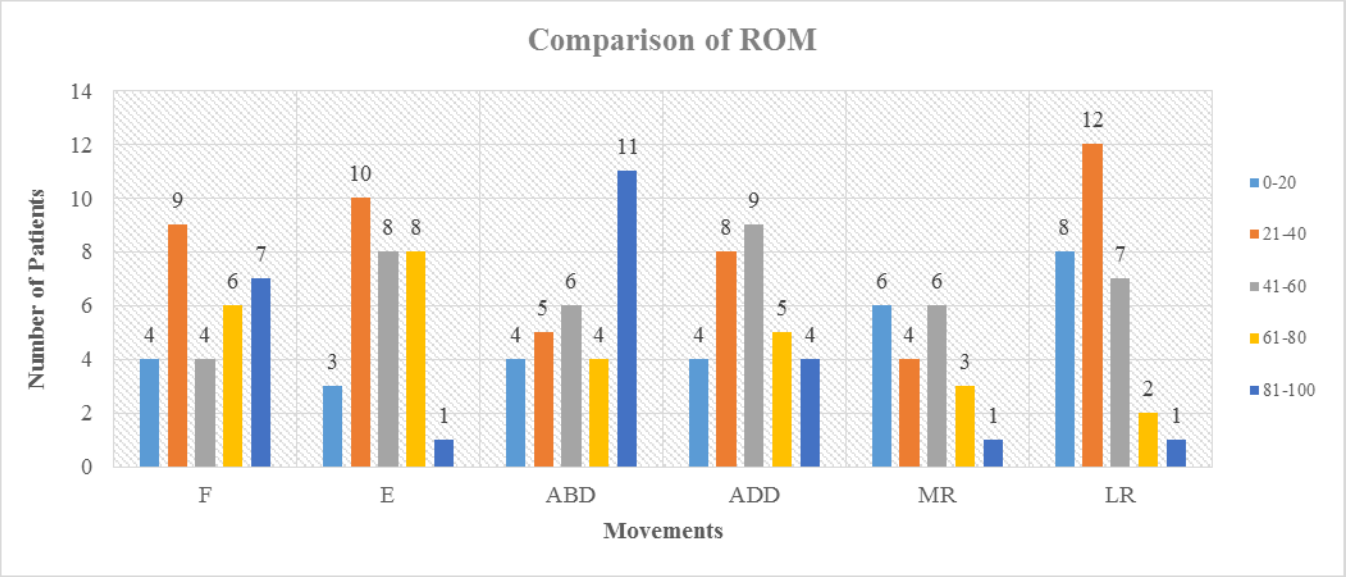
Graph 8- Distribution of data as er Leg length Discrepancy in cm.

8. Range Of Movement At Hip Joint

ROM of each movement at hip joint was measured with the help of a universal goniometer. The observed range was compared with normal range of movement at hip. Observed ranges of 6 different movemnts at hip joint compared with each other to understand severity of restriction. For this purpose percentage of movement respective to their normal values was obtained and grouped into 5 categories as 0-20%, 21-40%, 4160%, 61-80% and 81-100%.

Table 7: Frequency distribution Table of comparison of Movements.

ROM	0-20	21-40	41-60	61-80	81-100
Flexion (F)	4	9	4	6	7
Extension (E)	3	10	8	8	1
Abduction (ABD)	4	5	6	4	11
Adduction (ADD)	4	8	9	5	4
Medial Rotation (MR)	6	4	6	3	1
Lateral Rotation (LR)	8	12	7	2	1



Graph 9: Presentation of Comparison of Movements.

DISCUSSION

A) Discussion on Samprapti (Pathophysiology) of disease.

This century has witnessed many novel diseases with fatal nature and poor diagnoses. Many of these diseases have not been mentioned in the basic text of Ayurveda which suggest that these diseases may not have been in existence at that time and appeared later on. It has also been suggested that changing nature and environment with time were the chief contributors in the emergencies of new diseases.

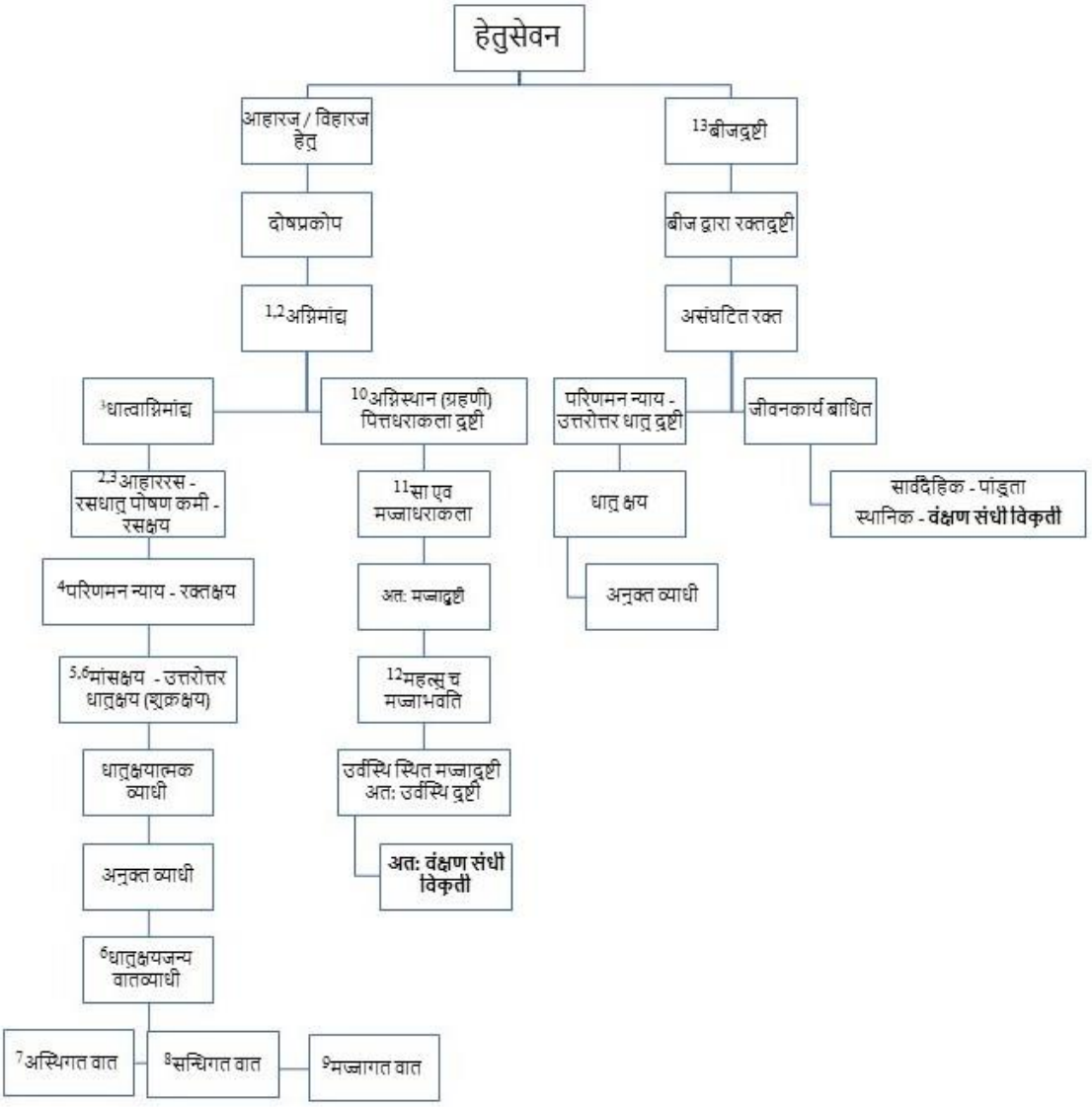
Modern sciences admit sickle cell anaemia as a genetic disorder and placed it in the incurable category. Sickle Cell Anaemia the term is not mentioned in Samhita it has to be treated as "**Anukta Vyadhi**" One of the great concept quoted by Acharya Sushruta.

Anukta Vyadhi-

नास्तिरोगोविनादोषैर्यस्मात्तस्माद्विचक्षण: Iअनुक्तानामपिदोषाणाम्लिंगैव्याधिमुपाचारेत्सु. सू.II 35/19II

Doshadushti is the basic cause of every disese. Since disease does not occur without doshas, the wise physician should recognize the symptoms of the vitiated doshas, even though not mentioned in ancient text book and should treat them. Following this Samrapti of SCA was extensively studied and along with doshadushti, beejadushti (genetical inheritance) was also studied as it is one of the cause mentioned in modern science.

Doshadushti-



Localized symptoms - *Jeevana Karma* is seen hampered on cellular level at *Vankshanan Sandhi* leading to destruction of bone cells which results in avascular necrosis which lead to *Sandhivikruti.*

In this study only *Vankshan Sandhi* is focused though other joints and bones are also involved. *Vankshan Sandhi* is the important joint of lower limb as it is the weight bearing joint, its derangement comparatively is more troublesome. One another reason for focusing hip joint is the frequency and severity of its (*Vankshana Sandhi)* derangement which is quite more than other joints.

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वातपूर्णदृतिस्पर्श: शोफंसंधिगातोSनिल:प्रसारणाकुंचनयोप्रवृत्तिंचसवेदनाIIअ.ह्.नि.15/14II

1) *Dhatukshayjanyavatavyadhi* particularly one of the three conditions-*Asthigatavata, Majjagatavata, Sandhigatavata.* In this study symptoms observed are indicative of one of these conditions.

2) Grahani is the location of Jatharagni and Pittadharakala. Hence with the Agni pittadharakala also gets vitiated. Pittadharakala is Majjadharakala. Kala is nothing but "कलाहिधात्वाशयान्तरमर्यादा". So if kala is vitiated then dhatu also gets vitiated. As mentioned by Acharya Sushruta, Majja is present in Sthoolasthi and Vankshan Sandhi is formed by Sthoolasthi. Hence vitiation of majjadharakala and Majjadhatu vitiates Asthi and thereby Sandhi. AdditionallyDhatukshaya leads to Vatavyadhi (Dhatukshayjanya) presenting with Sakthishoola, sandhishoola, neejabhagna of urvasthi etc.

B) Discussion on Observational Study –

In Sickle cell Dawakhana Dhadgaon approximately more than 4500 patients were under treatment

- In this study it is observed that all **30** patients were present with following complaints-

Krushangata (Emaciation)

Asthisoushirya (Osteoporosis)

Sandhishool (Joint pain)

Santatataruja (Continuous Pain)

Prasarana Akunchana Pravrutti Savedana.

- 2 patients were seen with *Pleehavruddhi and*

- 18 patients were observed *with Sakthishool*.

- Total 30 patients observed as per case record form. Data analyzed statistically. Results obtained are mentioned below.

o **Age**: In this study it was found that incidence was highest in the age groups 16-20 and 26-30 constituting 8 patients (26.66%) in each group. Followed by 6 patients (20%) in 21-25 age group, followed by 5 patients (16.66%) in 36-40 age group.

o **Gender**: Data showed that there were 17 male patients (56.66%) while female patients were 13(43.33%). Hence it can be said that data is nearly equally distributed slightly more towards male patients.

o **Side**: It was observed that 22 patients (73.33%) were complaining of unilateral involvement while 8 patients (26.66%) were bilaterally involved. Among unilateral involvement patients complaining for right side were 13(59.09%) more than that for left side 9(40.9%).

o **Pain**: When the pain scale was recorded for the sample population, it was observed that 6 patients (20%) experience severe pain, 16 patients (53.33%) experience moderate pain while 8 patients (26.66%) experience mild pain. All 30 patients presented with the complaint of pain.

o **Complaints**: Data distribution shows that all 30 patients were having pain, gait-lurching and difficulty in squatting. 5 patients had oedema at *Vankshana sandhi* and local temperature was raised in 3 patients only.

o **Leg length discrepancy** was observed in 19 patients (63%) while11 patients don't show any discrepancy. Out of which the data was arranged in 5 groups. Most of the patients i.e. 13 in this study undergo leg length shortening between 0.1 to 1 cm; 4 patients undergo 1.1-2cm difference; while 1 patient had 3cm and another one 5cm difference.

STATISTICAL ANALYSIS AND RESULTS

I. Leg length of right and left leg measured and compared with each other. Data analyzed using "**unpaired t-test".** t- value is 0.2022 and p-value is 0.8405. Since p-value is>0.05 difference between leg length of right and left leg was not found significant.

II. **Range of movement**: Range of movement of each movement at hip joint was measured with the help of universal goniometer. Observed range was compared with normal range.

- **Flexion**:As p-value is less than 0.05 observed range of flexion **proved significant**.

- **Extension**: As p-value is less than 0.05 observed range of extension **proved significant**.

- **Abduction**: As p-value is less than 0.05 observed range of abduction **proved significant**.

- **Adduction**: As p-value is less than 0.05 observed range of adduction **proved significant**.

- **Medial rotation**: As p-value is less than 0.05 observed range of medial rotation **proved significant**.

- **Lateral rotation**: As p-value is less than 0.05 observed range of lateral rotation **proved significant**.

III. **Radiological findings**: In this study it was found that **22** patients (73.33%) show radiological changes indicating **grade 4** avascular necrosis of hip. 1 patient show changes indicating grade 3 changes; 4 patients indicate grade 2 changes; while 3 patients are having normal radiographs. It implies the severity of necrosis of hip joint.

JJJ. **Comparison of range of movement of all movements at hip joint**

Observed ranges of 6 different movements at hip joint compared with each other to understand severity of restriction. Data analyzed statistically with the help of the **chi-square test** for association. Results obtained are- Chi-square value is 33.43 and P-value is 0.0302. As P-value is < 0.05 association between these movements proved significant.

Bone involvement is the commonest clinical manifestation of sickle cell disease both in the acute setting such as painful vaso-occlusive crises, and as a source of chronic progressive disability such as avascular necrosis. Management of these problems is often difficult because of the diagnostic imprecision of most laboratory and imaging investigations[8].

Radiological changes may lag 2 to 3 weeks behind the onset of clinical sign and symptoms[9]. If patient is diagnosed on clinical parameters only then the progression of the disease may be arrested to some extent as early diagnosis in these cases is of paramount importance.So the clinical symptoms observed during this study can be kept in mind to develop an assessment scale to arrest this alarming condition.

The treatment of choice for ONFH available today is hip replacement surgery. Hip replacement surgery for ONFH occurs at a young age, in comparison with the general population, and postoperative readmissions for painful vaso-occlusive crises are common. However, our findings of high postoperative readmissions rates for VOC is comparable to an older study of all-cause readmission rates for adults with SCD[10]and should not detract from the potential benefit of hip arthroplasty to decrease pain and improve mobility in SCD patients with systematic ONFH. As the SCD population ages,[11,12]epidemiology research should pivot toward assessing risk factors, defining morbidity and identifying effective therapies for chronic SCD complications such as ONFH.

One Cochrane Cystic Fibrosis and Genetic Disorders Group Haemoglobinopathies Trials Register search, comprising references identified from comprehensive electronic database searches and hand searches of relevant journals and abstract books of conference proceedings state that they found *no evidence* that adding hip core decompression to physical therapy achieves clinical improvement in people with sickle cell disease with avascular necrosis of bone compared to physical therapy alone.[13]

One study reveals that osteonecrosis is a revalent (30-50% by age 30) yet underdiagnosed complications in patients with sickle cell disease (SCD), commonly affecting the femora; and humeral head[14]. 25 studies(1 RCT, 1 case-control, 10 rospective and 13 retrospective

studies)were included from 544 unique citations(kapra= 0.82). A total of 2,460 participants with mean age of patients in each study between 19 and 40 years. Hip arthroplasty alone or in combination with non-surgical interventions were the most commonly used treatment options. Although hip arthroplasty offers post surgical pain relief, it is associated with high rates of revision. The paucity of RCT and the high risk of bias with the existing non-randomized studies highlight importance of RCT and the high risk of bias with the existing non-randomized studies highlight the importance of developing prospective controlled studies examining existing and novel therapies using a unified approach in determining the severity of ON and outcome measures[14].

This study focus on etiopathology from Ayurveda perspective so as to develop effective remedy to overcome the situation as well as to identify clinical symptoms for early diagnosis of the painful situation.

CONCLUSION:

➢ In this *Anukta Vyadhi* (Sickle Cell Anaemia) *Agnimandya* causing *Kshayatmaka Vikruti of dhatu* is observed. Consequences of this *Anukta Vyadhi*are observed in *Asthi* and *Sandhi*. At *Vankshan Sandhi symptoms of Sandhigat Vaata, Asthigata Vaata*or *Majjagatavaata (dhatukshayajanyavatavyadhi) were observed.*

➢ Structural changes in *vankshana Sandhi* in Sickle Cell Anemia Patients observed with the help of radiology are-

1. Sclerosis of head of femur.
2. Remodulation of femoral head.
3. Joint space narrowing with subarticular cyst formation.

➢ Clinical assessment criteria for hip joint pain in Sickle cell Anaemia patients is-

- Pain in groin, pain in hip joint.
- Lurching gait.
- Difficulty in squatting.
- Restricted/ arrested joint movement's especially Lateral rotation, Medial rotation, followed by Abduction.

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Nil

Conflict of interest:

There is no conflict of interest.

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