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TREATING SICKLE CELL ANEMIA CRISIS: ROLE OF IBUPROFENAND PARACETAMOL": RETROSPECTIVE STUDY

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Abstract: Sickle cell disease is a hereditary condition characterized by presence of HBS type of haemoglobin in red blood cells. In which red blood cells undergo morphological changes that create problems such as oxygen deficiency and vascular blood clots. The individuals are impacted because of a mutation in the hemoglobin ß chain. Hemoglobin S (HbS), a hemoglobin variation, is the cause of sickle cell disease (SCD). The crucial element is the substitution of valine for glutamic acid at position 6 in the β-globin chain. ***Materials and Methods****: A Retrospective study is conducted for 6 months in 230 sickle cell patients attending a tertiary care hospital in Telangana region. The data were collected from case sheets, patients and patient's representatives* ***Results:*** *In our study majority of patients belongs to age group 6 to 15 years was recorded to be152 patients and are from rural areas. In this study, type of DNA was found to be 59.56% homozygous and 40.43% are heterozygous & only 33.04% of the patient's both parents are traits. 50-70 is the maximum number of transfusions seen only in few patients.* ***Conclusion****:-Symptom relief was noted at different time points, with more rapid recovery in the tablet group than in the syrup group. We concluded that ibuprofen and paracetamol has good efficacy, well tolerated, safe and effective in reducing pain in sickle cell pain crisis.*

Key words: *Sickle cell anaemia, Ibuprofen, Paracetamol, Blood transfusions*

I. INTRODUCTION

Sickle Cell Disease in which red blood cells undergo morphological changes that create problems such as oxygen deficiency and vascular blood clots[1]. It is inherited disease with two abnormal sickle cell genes (one from each parent). This leads to the production of abnormal hemoglobin S, which causes red blood cells to become sickle- shaped, leading to various complications.[2]The condition brought on mutation in the hemoglobin genes, causes a change in the haemoglobin beta chain where valine is substituted for glutamate. This mutation leads to change in RBC shape Crises: Patients frequently endure excruciating periods referred to as "crises," which are associated with the sickled cells obstruction of blood flow.[3]

Painful Crises: When sickle cells blood supply is blocked, severe pain is experienced, usually in the chest, abdomen, joints, or bones. Due to sickle cell obstructions in the lungs, acute chest syndrome is a serious illness that causes fever, breathing difficulties, and chest pain.[4]

Risk factors include SCD genotypes (HbSS or HbSβ0-thalassemia), lower foetal haemoglobin (HbF) levels Higher steady-state hemoglobin and white blood cell counts History of asthma, atopy, and exposure tobacco[11].

vaso occulation: When sickle blood cells group together and stick to endothelial cells, they block tiny blood capillaries, leads to pain episodes, organ damages, and an increases the risk of infection. Elevated blood viscosity is caused by sickle-shaped red blood cells, as well as by larger concentrations of plasma proteins and inflammatory cells. This results in slower blood flow, which exacerbates vaso-occlusion (blood vessel blockage). **Inflammation and Oxidative Stress**: Haemolysis, the breakdown of red blood cells, releases iron and free heme, which exacerbates oxidative stress. In addition to activating endothelial cells and causing inflammation, this stress causes sickle cells to adhere to the blood vessel walls.

Endothelial Dysfunction: Sickle blood cells cause endothelial activation and dysfunction through their interactions with endothelial cells. Vaso-occlusion and tissue injury are made worse by the release of pro- inflammatory mediators, vasoconstriction, and a greater adherence of sickle cells to the endothelium.[5] **Diagnosis:** Hemoglobin electrophoresis is a test used to distinguish and identify various haemoglobin types.

•The sickle cell gene mutation can be accurately identified through genetic testing using sophisticated molecular techniques like PCR or gene sequencing.[3]

Treatment

Blood transfusions: Blood transfusions are essential for the treatment of sickle cell disease (SCD), but they carry a significant risk of major side effects. In order to treat acute complications such as splenic sequestration, transitory aplastic crisis, hyperhemolytic crisis, Acute chest syndrome (ACS), stroke, sickle hepatopathy, multisystem organ failure, and perioperative problems, intermittent red blood cell (RBC) transfusion is needed. Despite their advantages, these transfusions have the potential to cause both immediate and long-term negative effects.[6]

Pain Management: During painful episodes, treatment includes non-steroidal anti-inflammatory drugs (NSAIDs), opioids, and intravenous fluids to manage pain and prevent dehydration.[7]

IBUPROFEN & PARACETAMOL: The most widely used analgesic and antipyretic - to treat inflammatory diseases, mild to moderate pain.

Mechanism of action: It is a non-selective inhibitor of the enzymes cyclooxygenase-1 (COX-1) and cyclooxygenase-2 (COX-2), which are involved in the synthesis of prostaglandins, ibuprofen has anti- inflammatory properties.[8]

Counseling Points:

- Patient education is needed about the drugs like iron chelators, NSAIDs, vaccines and dietary approaches. Foods like beans, peas, cereals, pastas, green leafy vegetables increase the folate.
- Milk and yogurt increase the potassium levels and fish, nuts contain omega 3 fatty acids which reduce inflammation.
- Foods like bacon, ham, butter should avoid.
- Blood transfusion is necessary.
- Early detection of SCA is important before marriage to avoid trait.

II . METHODOLOGY

Study Procedure:

1. Patients are reviewed daily that are admitted in the sickle cell department.
2. All the necessary information collected from the patient case sheets, laboratory data, direct communication with patients and their care takers.

- **Study site :** Prathima Institute of Medical Sciences, Nagunoor, Karimnagar
- **Study Design :** Retrospective Observational study
- **Study Period :** 6 months
- **Sample size :** 230 Patients

❖ **STUDY ELIGIBILITY :**

Inclusion criteria :

•Children who visit Paediatric department with sickle cell anemia upto 18 years old in both In and out patients

Exclusion criteria : Above 18 years old. Children with thalassemia, sickle trait and sickle – thalassemia

III. RESULTS & DISCUSSION:

1. Age and Gender Wise Distribution

In our study 230 patients are admitted with sickle cell anemia with pain crisis. The age of participants varied from 1 to 18 years. In this 16.08% patients were admitted with age 1-5 years, 33.04% are admitted with age 6-10 years, 33.04% patients are admitted with age 11-15 years and 17.82% are admitted with 16-20 years of age. Male patients (65.2%) visited the ED more frequently than females (33.4%). 126 male patients (54.78%) and 104 female patients (45.21%) are visited. Male patients are more frequently admitted than female patients. A study conducted by **Hashim M. Taha *et al.*.**, they included 43 patients who made 270 emergency department visits. The ages of the participants varied from 5 to 18 years with a mean age of 12.1 years. In our study 230 patients are visited with SCA pain crisis. In this 16.08% pts visited at age 1-5 years, 33.04% are visited at age 6-10 years, 33.04% patients are visited at age 11-15 years and 17.82% are visited with 16-20 years of age. Male patients (65.2%) visited the ED more frequently than females (33.4%). 230 patients visited sickle cell department in our study. The age of participants varied from 1 to 18 years. 126 male patients (54.78%) and 104 female patients (45.21%) are visited. Males are mostly admitted than females.[9]

Table 1: Age and Gender Wise Distribution

AGE (YEARS)	MALE	MEAN	FEMALE	MEAN	P VALUE
1-5	15	31.5	22	26	0.19
6-10	48		28		
11-15	41		35		
16-20	22		19		

2. Area Wise Distribution

More members are from rural region, i.e., 143 pts (62.17%), remaining 87 pts were from Urban region (37.82%). A study conducted by **Joseph Telfair, Akhlaque Haque, Marc Etienne *et al.*.,** on Rural/urban differences in access to and utilization of services among people in Alabama with SCD. 468 patients (71%) from urban areas and 194 patients (29%) are from rural area. In our study among 230 patients, 143 (62.17%) patients are admitted from rural area and 87 (37.82%) patients are admitted from urban area. Population at rural are most prone from sickle cell disease than urban.[10]

Based on type of DNA in our study 40.43% patients are suffering with heterozygous SCD and 59.56% patients are suffering with homozygous SCD.

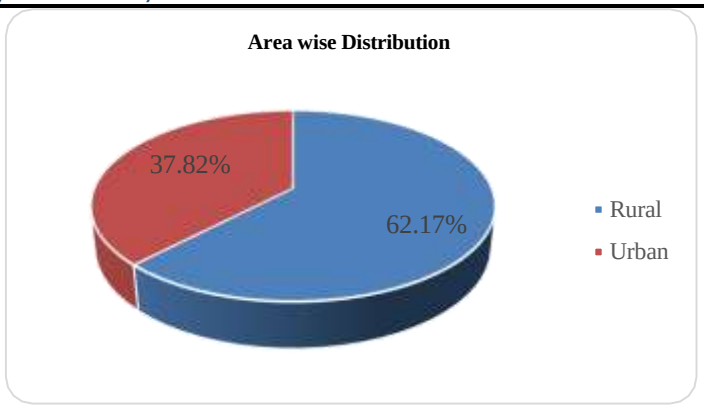

Figure 1: Area Wise Distribution

3. Family History

Based on family history 33.04% patients affected from mother and father, 28.26% patients from mother, 22.17% patients from father, 9.13% patients from grandfather, 5.65% from grandmother and 1.30% from both mother, father and siblings are affected as shown in the below figure 2.

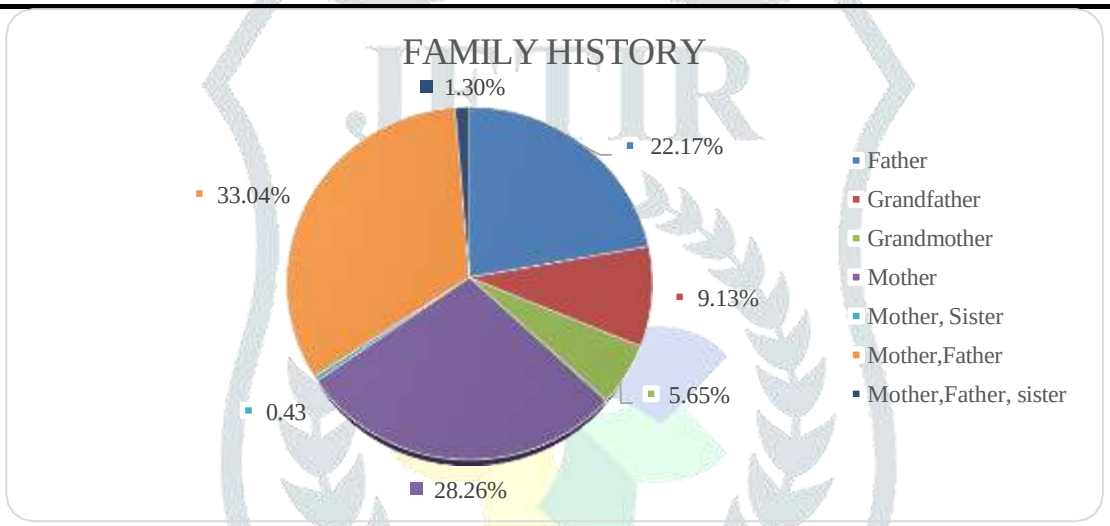

Figure 2: Family history

4. Distrbution of Sickel Cell Anaemia Patients Based on Symptoms

Among 230 Patients, most of the patients are having the symptoms of Shoulder Pain and Fever with 8.26% (19 Patients) as shown in theFigure 3 patients different symptoms.

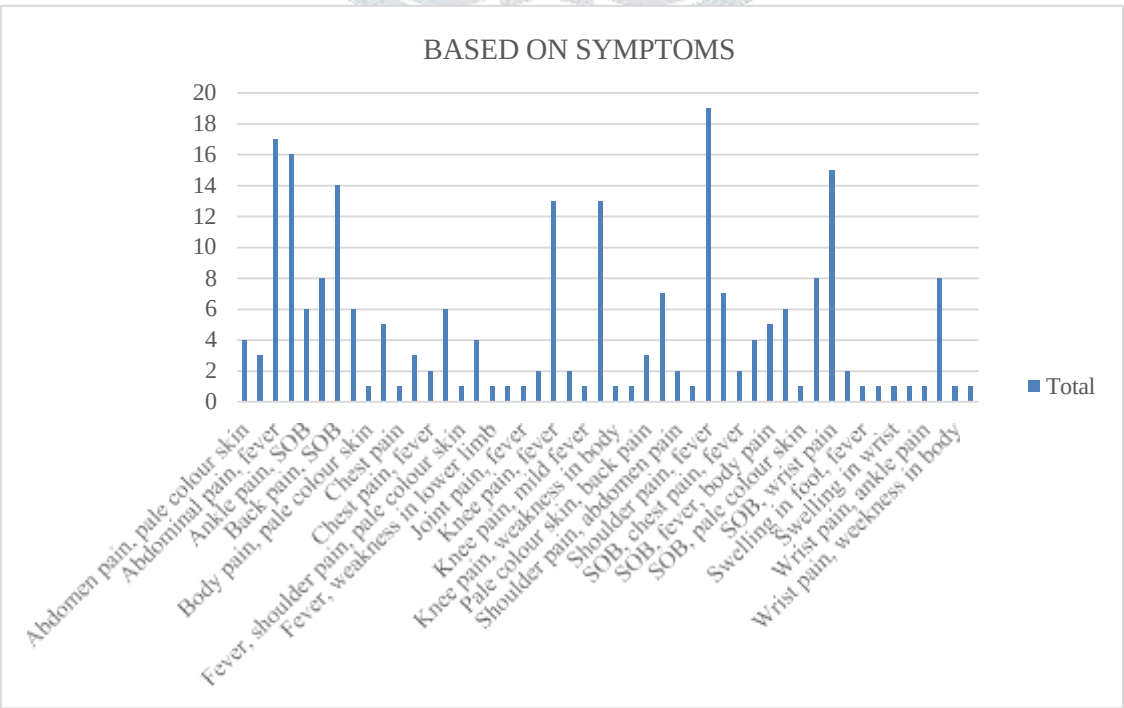

Figure 3: Based on the Symptoms of the study population

5. Number of Blood Transfusions Done In Sickle Cell Anaemia During Our Study

According to the study, among 230 patients majority of transfusions fell within the range of 1to 5 times seen in 63 patients followed by 35 patients underwent blood transfusions of 6 to 10 times and patients receiving a different number of transfusions as seen in Figure 4.

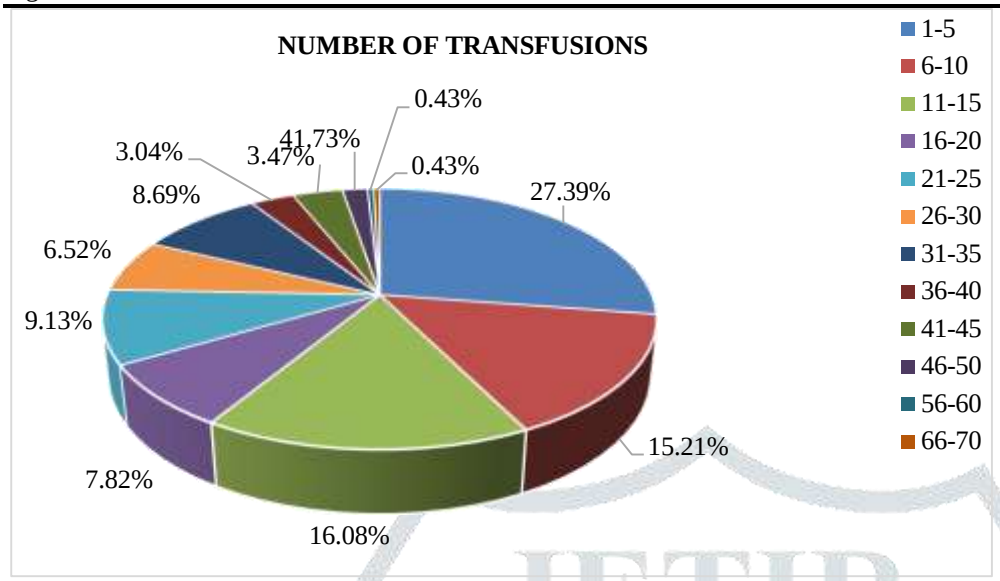

Figure 4: Number of Blood Transfusion among the study population

6. Symptoms Subsided with the Treatment

In our study, out of 230 patients 69 patients was prescribed with Tab. Ibuprofen and paracetamol twice a day and symptoms subsided to 20 patients (3.47%) at 8 hrs, 44 (5.65%) patients at 6hrs, 5 (1.73%) patients at 4hrs.

95 patients were given in Tab. Ibuprofen and paracetamol once a day and symptoms subsided to the 40 patients (17.39%) at 8 hrs, 49 (21.30%) patients at 6hrs, 6 (2.60%) patients at 4hrs.

25 patients were given in Syrup. Ibuprofen and paracetamol twice a day and symptoms subsided to the 8 patients (3.47%) at 8 hrs,13 (5.65%) patients at 6hrs, 4(1.73%) patients at 4hrs.

41 patients were given in Syrup. Ibuprofen and paracetamol once a day and symptoms subsided to the 13 patients (5.65%) at 8 hrs, 27 (11.73%) patients at 6hrs,1 (0.43%) patients at 4hrs.

Table 2: Treatment, Frequency and Symptoms Subsided Wise Distribution

Medication (Ibuprofen 400mg+Paracetamol 325mg)	Symptoms subsided at interval (Time)	Number of patients subsided symptoms with twice a day	PERCENTAGE (%)	Number of patients subsided symptoms with once a day	PERCENTAGE (%)
Syrup. Ibuprofen and Paracetamol	4hrs	4	1.73%	1	0.43%
Syrup. Ibuprofen and Paracetamol	6hrs	13	5.65%	27	11.73%
Syrup. Ibuprofen and Paracetamol	8hrs	8	3.47%	13	5.65%
Tab. Ibuprofen and paracetamol	4hrs	5	2.17%	6	2.60%
Tab. Ibuprofen and paracetamol	6hrs	44	19.13%	49	21.30%
Tab. Ibuprofen and paracetamol	8hrs	20	8.69%	40	17.39%

IV. CONCLUSION:

A study involving 230 patients aged 1-18 years from the sickle cell department showed a higher admission rate of male patients (54.78%, n=126) compared to female patients. In this study family history, 33.04% of patients had both parents affected, while

28.26% had mothers and 22.17% had fathers with this condition.

In the study we observed that, patients were prescribed either syrup (n=66) containing ibuprofen and paracetamol or tablets (n=164). Notably, the tablet group showed more rapid symptom relief compared to the syrup group at various time points.

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