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Original Research Article

EDUCATION OF PARENTS OF CHILDREN WITH SICKLE CELL DISEASE TO FACILITATE EARLY DIAGNOSIS OF CRISIS.

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Abstract:

Background: Sickle cell disease (SCD) is a haemoglobinopathy that affects millions throughout the world. It leads to poor quality of the life and increased mortality in children and young adults. In Vidarbha, SCD is common and it is pathetic to see children in morbid state dying due to some or the other crisis. Parents must be educated about the nature of the disease, earliest signs of crises and seek help, treat all febrile illnesses promptly. We contemplated that the key to success in the management of such children is parental education and this prompted us to undertake the present study.

Methodology:

Study Design: Interventional study.

Settings: Pediatric SCD clinic.

Study Duration: 1year

Data Collection: Parent's pre existing knowledge and awareness about sickle cell disease was assessed with the help of questionnaire and Objective Structured Clinical Examination (OSCE). They were then educated with the help of educational module comprising of pamphlets in Marathi, pictures, immunization protocols and also trained in clinical examination of their child to detect pallor, fever, jaundice, respiratory distress and splenic enlargement. After intervention post test and OSCE was conducted again.

Results: The pre-existing knowledge about inheritance, signs and symptoms of sickle cell anemia was high, but parents showed lack in skills of assessment of crises. These skills improved significantly after intervention. There was statistically significant ($p<0.05$) difference between average pre test and post test scores of each skill tested.

Conclusion: Modular teaching using OSCE helps in improving skills of parents for early detection of sickle cell crises.

Key Words: Sickle cell crises, Education of parents, OSCE.

Introduction:

Sickle-cell disease is characterized by red blood cells that assume an abnormal, rigid, sickle shape. Sickling decreases the cells' flexibility and results in a risk of various complications. The sickling occurs because of a mutation in the hemoglobin gene. [1] Mutations occur in the ß chain of Hemoglobin whereby the hydrophobic amino acid Valine takes the place of hydrophilic Glutamic acid at the sixth amino acid position of the polypeptide chain. This substitution creates a hydrophobic spot on the outside of the protein structure that sticks to the hydrophobic region of an adjacent haemoglobin molecule's beta chain. This clumping together (polymerization) of Hb S molecules into rigid fibers causes the "sickling" of red blood cells.[2]

The term "sickle cell crisis" is used to describe several independent acute conditions occurring in patients with sickle cell disease .Sickle cell crises include vaso-occlusive, aplastic, sequestration, hemolytic crises. Most episodes of sickle cell crises last between five and seven days. The commonest of these crises is the Vaso-occlusive. [3] Sickle cell disease is a haemoglobinopathy that affects millions throughout the world. Worldwide, approximately 300,000 infants are born with SCD.[4] According to a research by **Jaffer et al 2009**, knowledge about sickle cell disease and predisposing factors of sickle cell crises affect attitude (behaviors and beliefs) and therefore, parents/caretakers knowledge regarding sickle crises preventive measures is positively correlated with their attitude toward preventive practices.

Sickle cell anemia leads to poor quality of the life and increased mortality in children and young adults. The most affected group among children is those between six months to five years of age. This is the age at which the first sickle cell crisis usually appears. Parents must be educated about the nature of the disease. They must be able to recognize the earliest signs of crises and seek help, treat all febrile illnesses promptly. Parents should be instructed on how to palpate the abdomen to detect splenic enlargement, and the importance of observation for pallor, jaundice, and fever. We need to teach patients to seek medical care in certain situations, including persistent fever (>38.3oC), chest pain, shortness of breath, nausea, and vomiting, abdominal pain and persistent headache not experienced previously. Reinforcement should occur incrementally during the course of ongoing care. Family should be educated on the importance of hydration, diet, outpatient medications, and immunization protocol. In Vidarbha, SCD is common and it's pathetic to see children in morbid state dying due to some or the other crisis. Considering the high burden of sickle cell anaemia in Vidarbha region with reported prevalence in Wardha to be 5.7%[4] any effort towards reducing the morbidity and mortality is going to help patients.

Methodology:

Study Design: Prospective interventional study.

Settings: Pediatric SCD clinic at Acharya Vinoba Bhave Rural Hospital (AVBRH), Sawangi (Meghe), Wardha.

Study Duration: 1year

Study Participants: These were the parents of sickle cell anemia and their wards attending the sickle cell clinic at AVBRH. We included parents of patient with sickle cell trait also.

Exclusion Criteria:

Terminally ill patients of SCD.

Collection, presentation of data:

1. Parents of children with sickle cell disease, who gave consent for participating in this project and fulfill the inclusion criteria, formed the subjects of study. A predesigned and pre-validated questionnaire (containing both close ended and open

ended questions) was given to them to assess their pre-existing knowledge of various aspects of Sickle Cell Disease. The parents examined their child for fever, jaundice, pallor, respiratory rate, spleen. This constituted the pre test knowledge score and Objective Structured Clinical Examination (OSCE) score.

2. Through an educational module comprising of pictures depicting inheritance pattern, questionnaire, immunization protocols, facilities at sickle cell clinic at our Institute, they were educated about the disease. Parents were trained in clinical examination of their child for pallor, fever, jaundice, respiratory rate and palpation of abdomen to detect splenic enlargement with the help of simplified version of OSCE.

3. The same questionnaire was given as post test to them to assess their knowledge of various aspects of SCD and crises and simplified version of OSCE for pallor, fever, jaundice, respiratory rate and splenomegaly was arranged for them to assess their learning of ***"show how"*** domain of **Millers pyramid** which attributes to the competence of the parents.

4. The data was arranged in Microsoft Excel sheet and descriptive statistics was used to derive the percentages. The comparative analysis of the pretest and post tests was done using the paired't' test using SPSS software version 17.

Observations and Results:

In the present study, during study period total numbers of 38 parents were enrolled. Out of that, only 30 completed the study (8 parents withdrew during the post test phase).Out of these 30 parents, 24 had children with SS pattern and remaining six had AS pattern. Out of the 30 couples only one father was SS pattern. 3 (10%) of fathers had AA pattern while, 2 mothers were having normal electrophoresis pattern. Rest all were AS pattern (26 and 28 fathers and mothers respectively). Out of the 30 responders, 83.3% (25) had AS pattern and 16.6% were having AA pattern. Twenty two had male child affected and eight female child affected (2.7:1). We had 16 children in the age group of 0-5 years and 14 above 5 years.

Table:-1 , Frequency of sickle cell pattern in patients

TOTAL NO. OF PATIENTS	SICKLE CELL PATTERN	NUMBER OF PATIENTS	PERCENTAGE
30	"SS" PATTERN	24	80
	"AS" PATTERN	6	20

Table:2 Educational status of responder

EDUCATIONAL STATUS	RESPONDER	
	NUMBER	%
No formal education	1	3.33
Upto 5th standard	3	10
From 6th to 10th standard	12	40
11th and 12th standard	10	33.34
Graduation or more	4	13.33

Out of the 30 responder maximum 12(40%) parents had education between 6th to 10th standard followed by 10 (33.3%) parents who had education upto 12th standard. Only one parent was uneducated and 4 were graduate.

Table :-3 Pretest for questions 1, 2 & 3

QUESTION NUMBER	YES		NO		NOT SURE	
	NUMBER	%	NUMBER	%	NUMBER	%
1 (N = 30)	23	76.66	7	23.33	0	0
2 (N = 23)	5	21.74	4	17.4	14	60.86
3 (N=30)	30	100	0	0	0	0

In relation to questions about awareness of sickle cell anemia 23 (76.6%) were aware and 7 (23.3%) were unaware. Out of those who were aware of sickle cell only 5 (21.7%) knew exactly what is sickle cell anemia. 4 (17.4%) responded wrongly and 14 (60.8%) were not sure.

Table :-4 Pretest for questions 10, 11, 12, 13, 14, 15 and 16.

NO.	QUESTION	RESPONSES
1	Signs of a vaso-occlusive crisis?	- Swelling in body or joint. - Pain in whole body - Blood clot in vein - Weakness
2	Signs of aplastic crisis?	- Weakness - Giddiness - Blood transfusion - Prone to death
3	How will you look for pallor?	- Hb test - Has to go to doctor - Pale look - Eye look pale
4	How will you look for jaundice?	- Yellowish discolouration of eye. - Yellowish discolouration of urine. - Yellowish discolouration of body. - Blood will get reduced.
5	How will you check for fever?	- By touching the forehead - Increased heat of abdomen - Don't know - By thermometer
6	How will you count the respiratory rate?	- By pulse measurement - By seeing to and fro movement of abdomen - Don't know
	How do you check for splenomegaly?	- Don't know - By pressing the abdomen - Swelling on abdomen - Pain in abdomen

*(The responses are arranged in the order of frequency)

The table is showing the excerpts of responses to open ended questions

Only 13.3% parents were initially aware about vaccination but this increased to 40% post test and p value by chi square test was significant.

After providing education post test scores for question no 1, 2 and 3 (regarding the knowledge) improved significantly. 100% parents were aware about sickle cell and 86.66% had in depth knowledge about it.

Table:-5 Post test for questions 1, 2 & 3

QUESTION NUMBER	YES		NO		NOT SURE	
	NUMBER	%	NUMBER	%	NUMBER	%
1	30	100	0	0	0	0
2	26	86.66	4	13.33	0	0
3	30	100	0	0	0	0

There was also significant improvement in the responses to the open ended questions. As depicted in table

Table:-6 Post test for questions 10, 11, 12, 13, 14, 15 and 16.

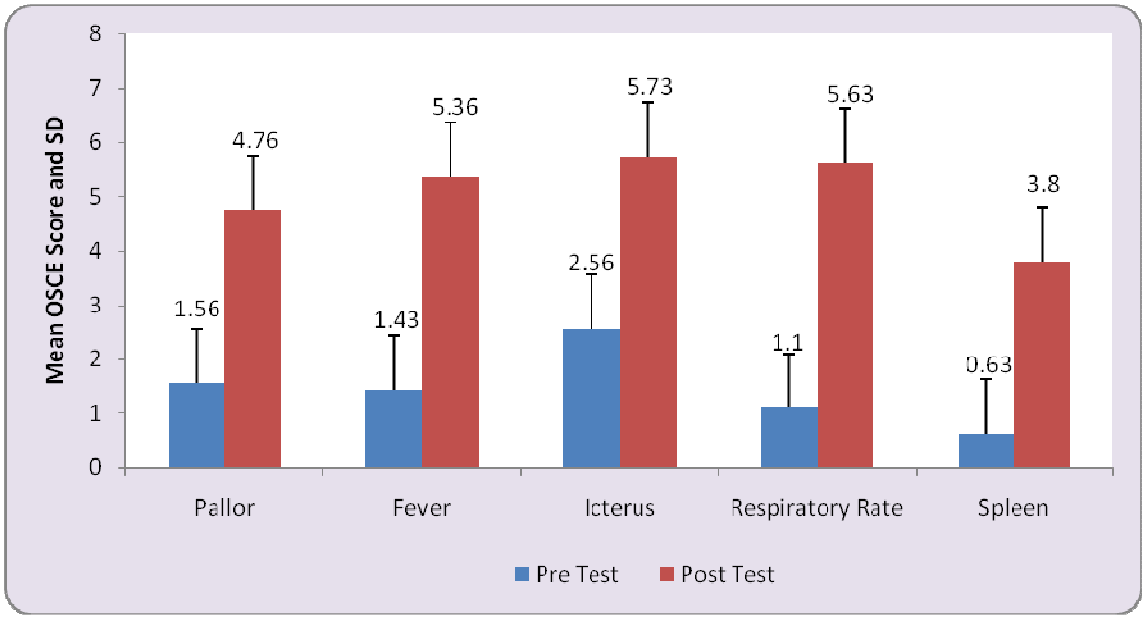
NO.	QUESTION	RESPONSES
1	Signs of a vaso-occlusive crisis?	- Pain in abdomen - Pain in joints - Swelling on hand and foot weakness
2	Signs of aplastic crisis?	- Weakness - Fever - Pallor - Requires blood transfusion
3	How will you look for pallor?	- Paleness in inner part of lower eye lid - Paleness on palm and nails - Paleness on tongue - Hb test
4	How will you look for jaundice?	- Yellowish discolouration of eye - Yellowish discolouration of urine - Yellowish discolouration of body
5	How will you check for fever?	- By thermometer - By touching the forehead with dorsal part of hand
6	How will you count the respiratory rate?	- In supine position seeing the abdomen for 1 min. - By seeing to and fro movement of abdomen
7	How do you check for splenomegaly?	- By pressing the abdomen - Don't know

Table:-7 Comparison of pre and post test OSCE score, (Paired t - test)

OSCE	Test	Mean	N	Std. Deviation	Std. Error Mean	Z-value	p-value
Pallor	Pre Test	1.56	30	1.10	0.20	12.30	0.000 S, p<0.05
	Post Test	4.76	30	1.19	0.21		
Fever	Pre Test	1.43	30	0.77	0.14	15.80	0.000 S, p<0.05
	Post Test	5.36	30	1.47	0.26		
Icterus	Pre Test	2.56	30	1.00	0.18	14.73	0.000 S, p<0.05
	Post Test	5.73	30	1.36	0.24		
Respiratory Rate	Pre Test	1.10	30	1.26	0.23	13.40	0.000 S, p<0.05
	Post Test	5.63	30	1.69	0.30		
Spleen	Pre Test	0.63	30	1.29	0.23	11.48	0.000 S, p<0.05
	Post Test	3.80	30	1.78	0.32		

As can be seen from the above table the average pretest score of OSCE for each station was less than the post test score and the calculated p value is < 0.0001 which is statically significant.

GRAPH:- 1



Discussion:

The present study was undertaken with aim of education of parents of children with sickle cell disease to facilitate early diagnosis of crises.

In the present study, out of the 30 parents involved 24 had a child with SS pattern.

Remaining 6 were AS pattern. In the electrophoresis pattern of the responder, out of the 30 responders, 83.3 per cent (25) had AS pattern and 16.6 per cent were having AA pattern. In this study 30 couples 50per cent mothers had education upto high school,40 per cent father were educated up to higher secondary school. Only one couple was uneducated.

As mentioned above, out of a total of 38 couples, eight couples withdrew from the post test. Out of these 8 couples, six had female child and two had male child and also in our study there were 22 male participants and rest females further showing that the parents have more predilection for their male children. According to a study done in Niger Delta University, Nigeria(2011) the study showed that sex correlates with the attitude of the parents towards the children with sickle cell disease although most of the participants were females because they interacted with the health facilities since mostly mothers stay with their children in the hospitals or accompany them to the hospitals, most mothers are more caring than fathers regardless of the diagnoses of their children even the prognosis.

In 30 responders maximum 12(40%) parents had education between 6^{th} to 10^{th} standard followed by 10 (33.3%) parents who had education upto 12^{th} standard. Only one parent was uneducated and 4 were graduate.

Studies done in Vidarbha area[6,7] also found lower class involvement in both disease and traits **M Sadat-Ali et al** [5]found in their study that socio-economic status does not influence the outcome in patients with sickle cell disease with respect to pattern and severity of the disease

In this study 24 (80 %) parents had non consanguineous marriage and only 6 (20%) had consanguineous marriage. Patients with chronic illnesses in sickle cell disease, their parents experience the same level of perceived illness intrusiveness as with other chronic diseases like end stage renal disease and vaso occlusive crisis as reported by **Devins**, 2010.

According to a study done in **Cleveland, Ohio** (2007) quantitative data methods show improvement of sickle cell disease management with Questionaire and suggested that in addition to using traditional approaches to engaging a designated caregiver in disease education and management, educational approaches need to include the children, who are critical agents in their own health care and disease management.

The percentage of consanginous marriage in sickle cell anaemia families is variable according to geographic area. The study by **Kamble and Chaturvedi** conducted nearby this study area, the percentage of consanginous marriage was 7 per cent which is similar to this study[8]

An average of about 30 per cent is seen in most Arab countries, through the prevalence of consanguinity range from about 25 per cent in Beirut [9] to 60 percent in Saudi Arabia and 90 per cent in some Bedouin communities in Kuwait and Saudi Arabia[10,11]

The awareness about sickle cell anemia is an important factor to prevent various crises. 76 per cent responders were aware and only 5 per cent new exactly what is sickle cell anemia. This was concluded after conducting pre test in this study.

There was improvement in the knowledge after the intervention. The general awareness about sickle cell anemia was seen in all parents while in depth knowledge was observed in 86 per cent. The parents also realized, the importance of vaccination in sickle cell anaemia. 40 per cent parents understood vaccination is necessary in patients of sickle cell anemia. In the questionnaire included in this study,

parents were asked both open and closed ended questions. After giving information on sickle cell anemia, most of the answers to the closed ended questions were correct. The qualities of responses to open ended questions were improved considerably and the accuracy increased.

The responders had no or poor skills for assessment of sickle cell anemia. Their judgment and physical assessment of signs seen in sickle cell anemia and crises improved.

Physical assessment of signs was checked through objective structured clinical examination

As can be seen in the results, average pretest score of OSCE for each station was less than the post test score and the calculated p value is < 0.0001 which is statically significant.

It was shown in many studies that the preexisting knowledge of the parents of children of sickle cell anemia improved [12,13,14,and 15]

Limitations of the study:

A study with large sample size can provide much more evidence for parental intervention in management of sickle cell anemia.

Summary :

In the present study, parents of children suffering from sickle cell anemia were selected. A pretest was prepared to test their knowledge and skills related with management of sickle cell anemia. The parents then went through the various modes of teaching to explain and understand the disease process. The knowledge of sickle cell anaemia and clinical skills assessed by using observed structured clinical examination method. The results were compared with the post test.

There was increase in the knowledge of all parents about sickle cell disease.

About 86% of the parents understood in depth idea of the disease.

The response to the questions about sickle cell anemia improved considerably. The parents used proper and precise words for describing the signs and symptoms of crises. The answers to the questions which were open type were complete and elaborative.

The acquisition of skills like assessment of pallor, icterus, fever, respiratory rate and splenomegaly was significant.

Conclusion:

- The crises in sickle cell anemia are prevented by regular administration of zinc, folic acid, immunization and proper follow up care. Parental awareness, skills in assessing their child helps in early intervention.
- The sickle cell anemia patients are at lifetime risk of crises. The crises are precipitated by minor illness. So, early recognition is priority.
- The study has stressed the role of parents in the management of sickle cell anemia. The attacks of crises if prevented will reduce the morbidity and mortality associated with sickle cell anemia.

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