

ABSTRACT

Sickle cell disease is a global health problem. It is an inherited blood disorder affecting the red blood cells. There are different types of sickle cell disease. Homozygous sickle cell (SS) disease, results when the infant inherits two abnormal sickle genes, one from each parent. In Jamaica SS disease affects approximately 1 in 300 births and some form of sickle cell disease, 1 in 150 births.

Complications of sickle cell disease include acute splenic sequestration. The cause and triggers of acute splenic sequestration (ASS) are largely unknown. The spleen becomes acutely enlarged, trapping a proportion of the red cell mass and leading to acute anaemia, circulatory failure and sometimes death. Emergency blood transfusion is extremely important in the management of acute splenic sequestration.

Parental education may allow prevention of death from ASS by early transfusion. Parents need to be taught how to diagnose splenic enlargement, and to recognize the signs and symptoms of increasing anaemia characterized by pallor. It is important for mothers to examine splenic size on a regular basis and when the child looks ill. At the Jamaican Sickle Cell Unit, parents are instructed to examine the spleen at least once a day.

The aim was to identify the parents' and guardians' knowledge of ASS, their attitudes towards the examination of the abdomen and practices in detecting an enlarging spleen. The objectives were to determine the percentage of parents and guardians who regularly examined the abdomen to determine spleen size; who had detected an enlarging spleen and the outcome; who were aware of the proper technique for examining the spleen; who were aware of what to do if an enlarging spleen was found; and identify the barriers that prevent regular examination for splenic enlargement.

The study was carried out on the parents and guardians of children four years and younger, with homozygous sickle cell disease, attending the sickle cell unit. At the time of the study, the population of children four years and younger with SS disease was 237. A questionnaire was designed based on the objectives of the study, to solicit data from the participants. The researcher administered the questionnaire to 60 parents and guardians who attended the clinic over a two-month period – March and April 2000. The participants were also asked to demonstrate their competence in examining the spleen.

One hundred percent of the parents and guardians interviewed, said that they had heard of the spleen and knew where the spleen was located. Although the parents and guardians were aware that they should examine the spleen everyday, only 50 percent did this. The other participants' examinations ranged

from twice a week to once per month. Fifty-five per cent of the participants said that the consequence of an enlarging spleen was death.

Twelve parents admitted to having found an enlarging spleen. Of these, 42 per cent said that they had taken the children to the Sickie Cell Unit immediately; 17 per cent had taken the children to the Sickie Cell Unit the following day; 25 per cent had taken the children to the hospital or a private doctor and 17 per cent said they did not take the children anywhere.

Thirty-six per cent of participants had a technique considered to be optimal in detecting an enlarging spleen. However, all 21 individuals began palpating from the left iliac fossa as opposed to the right. There was a significant association between 'attitude score' and 'regularity of abdominal palpation'.