Conclusion

Only one CDR developed for children with FN met all methodological standards and reached the highest level of evidence.

Background

Hereditary spherocytosis (HS) is a common inherited disorder that is characterised by anaemia, jaundice, and splenomegaly. Clinical severity is variable with most patients having a well-compensated hemolytic anaemia. The primary lesion in HS is loss of membrane surface area, leading to reduced deformability due to defects in the membrane proteins ankyrin, band 3, beta spectrin, alpha spectrin, or protein 4.2. Many isolated mutations have been identified in the genes encoding these membrane proteins; common hereditary spherocytosis-associated mutations have not been identified. The classic laboratory features of HS include minimal or no anaemia, reticulocytosis, an increased mean corpuscular haemoglobin concentration (MCHC), spherocytes on the peripheral blood smear, hyperbilirubinemia, and abnormal results on the osmotic fragility test.

Aim

Of the study is to evaluate the role of MCV, MCHC as a screen test to diagnose spherocytosis.

Methods

In our study are included 60 subjects, 30 children with HS and 30 children-control groups. Our patients with anaemia, jaundice, and splenomegaly are diagnose with HS by incubated osmotic fragility test, performed after incubating RBCs for 18–24 h under sterile conditions at 37°C.

Results

We found that 25% of pts. have mild HS, 20% moderate HS, 30% moderate to severe HS and 25% severe HS. In peripheral blood smear 7% of pts. had 0–5 spherocytes for field, 30% had 5–10 spherocytes for field and 63% had 10–15 spherocytes for field. 70% of pts. With HS have MCHC > 38%.

There is a positive correlation between MCHC and spherocytes in peripheral blood smear (p = 0.898, p < 0.001) and RDW (p = 0.647, p < 0.001), negative correlation between MCHC and MCV (r = -0.437 p < 0.001)

Conclusion

The dedication of hiperdense erythrociyte today is used as a new tool in diagnosing HS. The determination of MCHC constantly growing with other red cell index, MCV < 80 fl, RDW > 15 obtained from an electronic cell counter usually is enough to suggest for HS.

Key Words
Spherocytosis, MCHC, anaemia, children.

PS-090
HEREDITARY SPHEROCYTOSIS AND RED CELL INDICES MCHC, MCV, RDW

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10.1136/archdischild-2014-307384.384

Background

Acute lymphoblastic leukaemia (ALL) is the most common malignancy among children. Malnutrition remains a major concern for paediatric oncologists. Although studies have shown that malnutrition can negatively affect treatment outcome, results are still controversial. This retrospective cohort study aims at determining the prevalence of malnutrition and its association with treatment outcome and infection among children with ALL treated at the Children Cancer Centre in Lebanon (CCCL).

Methods

108 children and adolescents diagnosed with ALL between April 2002 and May 2010 were enrolled in the study. Anthropometric data were collected from patient's medical record upon diagnosis, at 3 and 6 months, and at the end of treatment. Body mass index (BMI) was calculated for children ≥ 2 years while weight for height ratio was used for patients < 2 years. Patients were considered underweight, stunted, or wasted if their z-scores were <-2SD.

Results

The prevalence of malnourished children was 27% at diagnosis and remained almost the same at the end of treatment. The odds ratio of having worse outcome in terms of relapse or death was higher among malnourished children with OR = 2.09, 95% CI = 0.3–13.4 and OR = 1.25 and 95% CI = 0.2–6.9 for death and relapse respectively. However this trend was