PEDIATRIK HEMATOLOGY
HEMATOLOGY – GENERAL

PP 56

Health-related quality of life for children with leukemia: child and parental perceptions

E. Ocak[1,*], A. Koca Yozgat[2], D. Kaçar[2], I. Ayrancı Sucaklı[1], N. Ozbek[2], O. Şükran Üneri[3], H. Yaralı[2]

[1] *Ankara City Hospital Children's Hospital, Department of Pediatrics, Ankara, Turkey*
[2] *Ankara City Hospital Children's Hospital, Department of Pediatric Hematology, Ankara, Turkey*
[3] *Ankara City Hospital, Children's Hospital, Department of Child and Adolescent Psychiatry, Ankara, Turkey*

Objective: The importance of health-related quality of life (HRQoL) in patients with acute lymphoblastic leukemia (ALL) has increased in recent years. This study aimed to assess HRQoL in children with ALL, affecting factors, and the relationship between parent proxy-report and child self-report HRQoL.

Methodology: The study sample consisted of 2–12 years old children with ALL between November 2016 and May 2017 at the University of Health Sciences Ankara Child Health and Diseases Hematology and Oncology Training and Research Hospital, Department of Pediatric Hematology. Patients and their parents (both mother and father) were enrolled in this cross-sectional study. Turkish version of the Pediatric Quality of Life Inventory (PedsQoLTM) 3.0 Cancer Modules were used to determine HRQoL. Patients' diagnosis, risk group according to the ALL-IC BFM 2009 protocol [standard risk group (SR), intermediate-risk group (IR), high-risk group (HR)], treatment status, the period between the cessation of the chemotherapy to the study and total hospitalization period, was obtained from the patients' medical record. Demographic data regarding the information on parents' age, education level, employment status, monthly income, and chronic medical condition were noted. Cardiovascular diseases, cancer, asthma, diabetes mellitus, thyroid disorders, and psychiatric problems were classified as chronic medical conditions by the Centers for Disease Control.

Results: A total of 59 patients (52,5% male) with a mean age of 7.28 ± 2.67 years at study period and 4.02 ± 2.51 years at diagnosis were enrolled. 57 patients (96.6%) were pre-B ALL and two (3.4%) patients were T-ALL. According to the risk groups; 18 (30.5%) patients had SRG, 25 (42.4%) patients had MRG and 16 (27.1%) patients had HRG. There were not any significant differences between on-treatment and off-treatment groups, age at study period, age at diagnosis, gender. There was no significant relationship between total scores of PedsQL cancer module self-report and the leukemia or sociodemographic features. According to subscales of self-report form; nausea and operational anxiety scores differed significantly by the treatment status; communication score varied considerably by the total hospitalization period; pain and hurt, cognitive problems and perceived physical appearance scores differed significantly by maternal chronic disease status ($p < 0.05$). No significant relationship was found between the total scores of the PedsQoLTM-cancer module parent-proxy report (father) and leukemia or sociodemographic features. The presence of maternal chronic disease was significantly related to the total score of the PedsQLTM-cancer module parent-proxy report (mother) ($p < 0.05$). There was a moderate correlation between total scores of child and mother ($p < 0.05$, $r = 0.419$) but not with the father.

Conclusion: Children on-treatment had significant problems in nausea and procedural anxiety subscales; however, children who hospitalized more had fewer issues in the communication subscale. Also, children whose mother had chronic disease had poorer HRQoL regarding pain and hurt cognitive problems, and treatment anxiety. Given the importance of assessment and monitoring HRQoL in children with ALL, health professionals should be aware of how parents' chronic disease affects HRQoL. Psychosocial support should be provided to children and their parents, especially whose parents have a chronic illness.

https://doi.org/10.1016/j.htct.2020.09.119

PP 57

Rare infectious agents in children with hematological disease

Y. Akcabelen[*], A. Koca Yozgat, Z. Guzelkucuk, O. Arman Bilir, M. Isik, D. Kacar, D. Gurlek Gokcebay, N. Yarali

Ministry of Health Ankara City Hospital, Ankara, Turkey

Objective: Infections are among the most important causes of mortality and morbidity in immunocompromised children. Although, the microbiological agents are usually opportunistic infections, sometimes rare infectious agents can also cause severe clinical conditions. Here, we present eight different microbial agents that can rarely cause infections in children with febrile neutropenia.

Case report: Case 1 is a 5-year-old girl with acute lymphoblastic leukemia (ALL) had a bloodstream infection during the reinduction therapy. Candida pelliculosa was detected in the blood culture taken from the port catheter. The catheter was removed and the patient was successfully treated with caspofungin. Case 2 is a 1-year-old girl with acute myeloblastic leukemia had a bloodstream infection during the first induction therapy. Cronobacter sakazakii was detected in peripheral blood culture. The patient was treated with cefepime and amikacin without port removal. Case 3 is a 5 month old girl with hemophagocytic lymphohistiocytosis had a pneumonia during the HLH 2004 protocol. Nocardia asteroides was detected in the bronchoalveolar lavage fluid. The patient was treated with trimethoprim-sulfamethoxazole and meropenem, however, she died of sepsis and multiple organ failure. Case 4 is a 2-year-old girl with ALL had a sepsis during the consolidation therapy. Candida tropicalis was detected in the port catheter and peripheral blood culture and renal abscess had developed. The patient was treated with broad spectrum antibiotics however she died sepsis and multiple

organ failure. Case 5 is a 9-year-old male with ALL had a bloodstream and port catheter infection after the first induction therapy. Herbaspirillum huttiense was detected in the blood culture taken from the port catheter. The patient was successfully treated with meropenem without port removal. Case 6 is a 10-year-old girl with ALL had a bloodstream and port catheter infection during the second induction therapy. Ralstonia pickettii was detected in the blood culture taken from the port catheter. The catheter was removed and the patient was successfully treated with piperacillin-tazobactam. Case 7 is a 7 month old male with Juvenile myelomonocytic leukemia had a bloodstream and port catheter infection in the neutropenic period. The patient was constantly inserting the port catheter into her mouth. Staphylococcus salivarius was detected in the blood culture taken from the port catheter. Then, 5 day after, Rothia mucilaginosa was detected in the peripheral blood culture. The patient was successfully treated with meropenem without port removal. Case 8 is a 9-year-old girl with ALL had a infective endocarditis and sepsis during the induction therapy. Magnusiomyces capitatus was detected in the peripheral blood culture. The patient was treated with fluconazole and amphotericin-B, but she died of multi-organ failure.

Conclusion: Many different microorganisms can cause infections in immune-compromised children as a result of primary disease or chemotherapy. Though empiric antibiotic therapy should be initiated early, the treatment should be revised according to the antibiogram and catheter should be removed as needed.

https://doi.org/10.1016/j.htct.2020.09.120

PP 58

Idiopathic hypereosinophilic syndrome associated pulmonary hypertension in a child

Y. Akcabelen*, T. Bayhan, G. Cinel, I. Ece, N. Ozbek

Ministry of Health Ankara City Hospital, Ankara, Turkey

Objective: Hypereosinophilic syndrom (HES) is defined by showing eosinophilic infiltration in any tissue or organ and increased eosinophils in peripheral blood. Other pathologies that cause eosinophil increase must be excluded. Pulmonary eosinophilic infiltration may have different symptoms and signs, but clinical presentation as PHT has not been shown in children.

Case report: A 6-month-old girl presented with dyspnea and hypoxia. A blood cell count and a morphological evaluation of a peripheral blood smear and confirmed hypereosinophilia (white blood cells 40,600/μL, eosinophils 18,900/μL, hemoglobin 10.3 g/dL, and platelets 425,000/μL). There was not any cellular morphological abnormalities in bone marrow aspiration examination. Pnomonia and parasites, allergic diseases, clonal abnormalities, cancer and vasculitis that might have caused HES were excluded. Echocardiogram showed 38 mmHg for pulmonary arterial pressure (PAP), suggesting pulmonary hypertension (PHT). After exclusion of other causes such as vasculitis, connective tissue diseases, bronchopulmonary displasia, congenital heart diseases, lung diseases, and chronic thromboembolic PHT. The patient was diagnosed with pulmonary arterial hypertension associated with idiopathic HES. Methylprednisolone treatment was started at 2 mg/kg/day. PHT and HES were both improved in the evaluation one month later.

Conclusion: Eosinophilic infiltration causes thickening and remodeling of the pulmonary artery intima and media, thereby causing pulmonary hypertension. Thus, PHT can be seen as HES clinical presentation. With corticosteroid therapy, HES and PHT clinical findings can be controlled.

https://doi.org/10.1016/j.htct.2020.09.121

PP 59

A rare variant of dyskeratosis congenita: RTEL1 defect

C. Coskun[1,*], S. Unal[2], N. Akarsu[2]

[1] *Hacettepe University, Department of Pediatric Hematology, Ankara, Turkey*
[2] *Hacettepe University, Research Center for Fanconi Anemia and Other Inherited Bone Marrow Failure Syndromes, Ankara, Turkey*

Objective: Dyskeratosis congenita (DC) is a rare hereditary disorder characterized by bone marrow failure, malignancy predisposition and skin findings. As the disease progresses, patients may develop pulmonary fibrosis, esophageal stenosis, urethral stenosis and liver cirrhosis. Herein, we present a patient who was referred with a diagnosis Diamond Blackfan anemia and was diagnosed to have dyskeratosis congenita on whole exome sequencing (WES).

Case report: A 18 month-old girl who was initially transfused at the age of three-months old and was on mothly transfusion programme, was referred to our center for molecular work-up with a diagnosis of DBA. There was second degree consanguinity between parents. On physical examination, body weight: 8.7 kg (5th percentile) height: 44 cm (<3rd percentile) was measured. Cubitus valgus was seen with camptodactyly. Liver and spleen were not palpable. Complete blood count showed hemoglobin (Hb) 7.9 g/dL, mean corpuscular volume (MCV) 104.1 fl, white blood count 6.9×10^9/L, absolute neutrophil count 1.3×10^9/L, platelet count 682×10^9/L, reticulocytes 2% and peripheral smear showed hypochromia and macrocytosis in erythrocytes. Biochemical parameters, globin electrophoresis, vitamin B12 and folic acid levels were normal. Parvovirus B19 was negative. ADA2 enzyme level was determined as 24 U/L (5–20 U/L). Steroid was started at the age of 18 month-old with a clinical suspicion of DBA. She became transfusion independent after steroid initiation. WES analysis for DBA from the patient revealed RTEL1 gene mutation (c.1368G> T p.1trp456Cys). This mutation was found compatible with DC and no other mutations in DBA related genes were detected, including CNV analyses for large deletions. Steroid was ceased gradually and she did not require further transfusions after complete cessation.

Results: In dyskeratosis congenita cases where the disease does not follow classical presentation, the use of genetic