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.

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Idiopathic hypereosinophilic syndrome associated pulmonary hypertension in a child

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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.

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PP 59

A rare variant of dyskeratosis congenita: RTEL1 defect

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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