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326 DRESS syndrome should be included in the differential diagnosis of life-threatening systemic symptoms in patients with cystic fibrosis

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A stable 20-year-old patient with CF (M, F508del/F508del, last FEV1: 92% pr) was admitted for sudden onset of high-grade fever, FEV1 drop (72% pr) without increased cough, diffuse pain and generalized lymphadenopathies. Initial check-up revealed the presence of prominent and very recent bilateral hilar lymphadenopathies along with a marked inflammatory syndrome. In 4 weeks, IgG level has risen from 15.7 to 30.4 g/L. Within 24 hours, the patient developed a generalized non-pruritic purpuric rash. Despite broad-spectrum IV AB (tobramycin and ceftazidime), daily fever persisted (up to 40°C). On the 6th day, the clinical picture worsened and became life-threatening with impaired renal function, multiple serositis, need for supplemental oxygen, hepatic cytolysis and coagulation abnormalities, major hypergammaglobulinemia (43.9 g/L) and marked hyper eosinophilia (4.870/μL). At this point, all non-essential drugs were interrupted with the exception of ceftazidime. Within 7 days, clinical, biological and radiological features all improved. The patient was released under his previous outpatient treatment and without precise diagnosis as all hematologic, autoimmune and viral detection tests were negative. He came back 2 days later due to the recurrence of malaise, fever, rash and biological abnormalities. Rifampicin and moxifloxacin were immediately interrupted and the picture resolved quickly. The patient then disclosed that he kept taking these drugs (no more prescribed) till day 6 of the hospitalization. We concluded that our patient suffered from a drug reaction with eosinophilia and systemic symptoms (DRESS) syndrome, rifampicin being the most likely causal agent.

327 Severe anemia in infancy may be the first sign of cystic fibrosis

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Objectives: Although cystic fibrosis is common, presentation may be atypical. Anemia in CF has been previously described with varying prevalence as high as 33%. To the best knowledge of the authors, there is no data about severe anemia as an initiation sign.

Methods: Patients with diagnosis as CF were reviewed in terms of anemia and hematologic abnormalities as the initiation sign.

Results: In 2009–2014, 62 patients were diagnosed as CF in Gazi University Pediatric Pulmonology Department. Hematologic abnormalities were the first symptom in 7 of 62 (11.2%) patients. The mean age was 3.3±1.1 months and 4 (57%) of them were male. None of them had any complaints about the respiratory system in time of diagnosis.

Severe anemia (hemoglobin <7 g/dl) was detected in all 7 patients. Mean hemoglobin level was 5.2±0.56 g/dl. Neither thrombocytopenia, thrombocytosis nor leukocytosis, leukopenia, lymphopenia were detected in any patient. In 2 patients international normalized ratio and prothrombin time were detected abnormal. Three of them had congestive heart failure together, and 6 patients required blood transfusion; 3 patients had the need for intensive care. Primarily, patients had been investigated for iron, folic acid or vitamin B12 deficiency. All of them had normal or even higher iron, ferritin, vitamin B12 and folic acid values. After the CF diagnosis, with vitamin E supplementation, anemia was recovered in all of the patients.

Conclusion: Severe anemia may be the first sign of cystic fibrosis especially in infancy. Cystic fibrosis should be included in the differential diagnosis of severe anemia of infancy.

328 Use of an insulin pump combined with the FreeStyle Libre interstitial glucose monitor in a needle-phobic adolescent with cystic fibrosis-related diabetes

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Cystic fibrosis-related diabetes (CFRD) occurs in approximately 20% of adolescents with CF. Self-measured blood glucose (SMBG) testing is required for optimal glycaemic control. Calorie supplementation makes diabetes management challenging. This has led to insulin pump therapy being recommended. Frequent SMBG testing can represent a significant hurdle to effective management.

Our patient is a 16-year-old girl with CF. She has had infections with *Pseudomonas*, *Aspergillus*, *Staphylococcus aureus* and *Mycobacterium abscessus*. She was diagnosed with CF-related diabetes aged 13 years. Her severe needle phobia made diabetes management extremely challenging. She refused to perform SMBGs or alter insulin therapy, resulting in uncontrolled hyperglycaemia [HbA1c 110 mmol/mol (12.2%)].

She trialled an insulin pump but refused to perform SMBGs, leading to sub-optimal glycaemic control [HbA1c 82 mmol/mol (9.6%)]. This prompted us to try the Freestyle Libre iGM (Abbott Diabetes Care, UK) combining insulin pump therapy and interstitial glucose monitoring. It relies on a subcutaneous glucose sensor, placed on the upper arm. Glucose levels are checked by “scanning” the sensor with a reader obviating the need for regular SMBG testing. It reports current glucose concentration, glucose trend and displays the previous 8 hours as a trend.

Use of the Freestyle Libre iGM was very successful in managing glucose levels [predicted HbA1c 50 mmol/mol (6.7%)]. It was acceptable to the patient and led to improved glycaemic control [HbA1c 67 mmol/mol (8.5%)].

This technology has the potential to greatly improve glycaemic control in CFRD, particularly in patients unwilling to perform SMBG measurements.

329 Partial recovery of exocrine pancreatic function and intestinal fat absorption after ivacaftor treatment

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Objectives: Ivacaftor treatment results in rapid improvement of lung function and weight gain. We hypothesize that the reported ivacaftor induced weight gain is related to recovery of exocrine pancreatic function and improved intestinal fat absorption.

Methods: We measured fecal elastase-1 (FE), FEV1 and body height and weight, in a 13-year old girl with CF (dF508/S1251N) with proven pancreatic insufficiency (FE <15 ug/g; maintenance PERT: ~2000 F.I.P.-units lipase per gram of dietary fat; CFA 98%) before and 3 months after the start of ivacaftor treatment. Additionally we measured the coefficient of fat absorption, without PERT, three months after the start of ivacaftor treatment.

Results: The patient reported a striking improvement in the general condition and body weight. FEV1 increased from 57% to 89%. Despite a decrease in the total dietary energy intake of 9%, the z-score of the BMI increased from 0.1 to 0.9 (+0.8SDS), after 3 months of ivacaftor treatment compared to baseline. FE increased to 54 ug/g. The CFA without PERT was 88%.

Conclusion: Our results, based on a single case study, suggest a partial recovery of exocrine pancreatic function, already, after 3 month of ivacaftor treatment. These findings are paralleled by a trend towards normalization of the spontaneous intestinal fat absorption. Noteworthy in this case is the increase in BMI, despite the still below normal CFA and decreased dietary intake. The latter suggests that, besides improved intestinal energy absorption, also lower total daily energy expenditure relates to the observed weight gain in CF patients during successful ivacaftor treatment.