

Posters

13. Case Presentations

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359 DIOS syndrome in 11-month-old infant with cystic fibrosisB. Kordys-Darmolińska¹, H. Wos², U. Grzybowska-Chlebowczyk², M. Dębowska³.¹Silesian Medical University, Department of Pediatrics, Katowice, Poland;²Medical Silesian University, Pediatric Department, Katowice, Poland;³Uppersilesian Child Health Centre, Pediatric Department, Katowice, Poland

Introduction: DIOS syndrome in patients with CF occurs with a frequency of 6.2/1000 patients/year, mainly in adolescents and young adults. It is extremely rare in younger children. Known risk factors for DIOS include: meconium obstructions in the neonatal period, abnormal enzyme supplementation, dietary errors, dehydration and others.

Objective: Analysis of the clinical assessment of risk factors for DIOS syndrome in an 11-month-old infant suffering from cystic fibrosis.

Material and Methods: We present a case of DIOS in an 11-month-old female infant diagnosed with CF by newborn screening test. The baby was referred to the district hospital because of febrile conditions, reduction in fluid consumption and eating, and ion disorders. On admission the child had features of dehydration and bloated belly with lazy motility. Additional examination revealed high rates of inflammation, electrolyte disturbances, hypoproteinaemia, and high rates of acidic steatocrit. Despite the persistence of observed treatment of infections parameters, growth of abdominal circumference, disposable vomiting. The diagnostic imaging studies, including the intestinal passage, showed retention of contrast around the ileocaecal bowel. Child consulted surgery. Oral feeding was stopped, starting rectal infusions of N-acetylcysteine, irrigation, included metronidazole, standard realimentation with enzyme supplementation. Improvement in clinical status was obtained.

Conclusions:

1. Avoidance and effective treatment of states of dehydration and the proper enzyme supplementation may reduce the risk for DIOS syndrome in infants.
2. Coexistence of several risk factors in the same child greatly increases the risk of developing ovarian DIOS.

360 A case series of thrombotic and thromboembolic complications of central venous access in cystic fibrosis patientsN. Kandamany¹, M. Tariq¹, B. Elnazir¹, P. Grealley¹. ¹Adelaide & Meath Hospital inc the National Children's Hospital, Tallaght, Respiratory, Dublin, Ireland

Objectives: To describe the diagnostic and treatment course of 4 cystic fibrosis patients with unusual and potentially fatal thrombotic and thromboembolic complications of indwelling central venous access systems.

Methods: Patient demographics, histories, biochemical data and imaging were compiled retrospectively through in-patient hospital documentation and hospital laboratory and radiological systems. Cardiology data was obtained from echocardiography reports and vascular data was obtained from pulmonary catheter laboratory reports.

Conclusions: This novel case series addresses several important diagnostic and therapeutic issues arising from the hypercoagulability of cystic fibrosis patients and their need for long-term venous access. In addition to discussing the potentially fatal complications of indwelling central venous access devices, this case series also highlights an emerging challenge arising from the improved life expectancy of cystic fibrosis patients today.