Journal of Cystic Fibrosis 16 (2017) S1

Journal of Cystic Fibrosis

www.elsevier.com/locate/jcf

Editorial

Introduction

Cystic fibrosis is an autosomal recessive, multisystem disease that affects the lungs, pancreas, intestines, hepatobiliary tract, and male reproductive tract and is caused by mutations in the cystic fibrosis transmembrane conductance regulator (*CFTR*) gene, which is located on chromosome band 7q31.2. Until the advent of pancreatic enzyme replacement therapy, patients with CF rarely lived past the first year of life. With this life saving therapy, survival for the first time, could be extended.

With advances in nutritional and respiratory care, the current median survival age ranges from 40.6 to 50.9 years in the US and Canada, respectively. The majority of children with CF are now expected to survive and transition to adult CF multidisciplinary health care teams, including adult gastroenterologists.

Although there has been a strong focus on the pulmonary aspects of CF, gastrointestinal (GI) manifestations play a significant role in CF beginning in childhood. Exocrine pancreatic insufficiency is present in the majority of patients with CF, often already at birth or in the first weeks of life. Many patients with CF suffer from intestinal fat malabsorption, leading to poor growth, difficulty gaining weight and deficiencies of fat-soluble vitamins and essential fatty acids. A high percentage of patients suffer from gastroesophageal reflux disease and intestinal dysmotility. A substantial number of patients with CF have intestinal obstruction phenomena varying from recurrent abdominal pain and constipation to the potentially life-threatening distal intestinal obstruction syndrome (DIOS) as well as meconium ileus. Finally, up to 25% of CF patients develop some form of CF related liver disease (CFLD). CFLD occurs in different levels of severity. The most severe form of CFLD is the development of cirrhosis and portal hypertension, which occurs in about 5% of the patients with CF.

Increased survival outcomes and the multisystem nature of the disease, including the involvement of hepatobiliary and gastrointestinal tracts, now require the need for more extensive knowledge and expertise in CF among gastroenterologists. Through the support of the U.S. CF Foundation, a two-pronged approach has been taken to 1) address the need for pediatric and adult gastroenterologists with expertise in the GI manifestations of CF and 2) define GI best practices. To address expansion of the GI physician workforce, in 2014, the CF Foundation launched a training program called DIGEST (Developing Innovative Gastrointestinal Training). This three-year on-the-job educational program focuses on training pediatric and adult gastroenterologists across the U.S. and now North America, in the management of the GI aspects of CF and includes monthly didactic and case based webinars as well as scholarly projects. The second aspect is the defining of best practices in the evaluation and treatment of the GI aspects of CF. As a result, we have commissioned 10 articles which have been put together by DIGEST awardees and senior faculty experts in GI and CF which will bring the reader up to date on 10 key topics. Although there is little evidence based research to guide evaluation and treatment, the following 10 articles represent our current thinking on the approach to evaluation and treatment of the following conditions: gastroesophageal reflux disease; gastrointestinal dysmotility, gastroparesis, and small intestinal bacterial overgrowth; chronic abdominal pain; meconium ileus; constipation and DIOS; CF related cirrhosis; gallstones and gallbladder disease; pancreatic insufficiency; pancreatitis; and prevention of nutritional failure. We envision that these articles will serve as a starting point for how we think about the diagnosis and treatment of the GI aspects of CF related disease and that over time, these will be modified and updated. In addition, these articles highlight the many gaps that exist and areas for future research.

Steven D. Freedman[1]

Division of Translational Research and The Pancreas Center, Beth Israel Deaconess Medical Center, Boston, MA, United States

Boston Children's Hospital, Division of Respiratory Diseases | Cystic Fibrosis Center, Boston, MA, United States

Corresponding author at: Beth Israel Deaconess Medical Center, 330 Brookline Ave., Dana 501, Boston, MA 02215, USA.

E-mail address: sfreedma@bidmc.harvard.edu.

[1]Dr. Freedman was supported in part through a grant from the Cystic Fibrosis Foundation and has no conflict of interest.

Michael Wilschanski[2]

Pediatric Gastroenterology, Hadassah University Hospitals, Mt. Scopus PO Box 24035, Jerusalem 91240, Israel

E-mail address: michaelwil@hadassah.org.il.

[2]Dr. M. Wilschanski was supported in part through a grant from the Cystic Fibrosis Foundation and has no conflict of interest.

http://dx.doi.org/10.1016/j.jcf.2017.07.013

1569-1993/© 2017 European Cystic Fibrosis Society. Published by Elsevier B.V. All rights reserved.