Iran J Pediatr. 2015 April; 25(2):e255. DOI: 10.5812/ijp.255

Published online 2015 April 18. **Letter**

Clinical Presentations of Cystic Fibrosis in Iranian Children

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Received: April 18, 2014; **Accepted:** May 14, 2014

Keywords: Cystic Fibrosis; Failure to thrive; Iran

Dear Editor,

Cystic Fibrosis (CF), characterized by abnormal transport of chloride and sodium ion across the epithelium, leads to thick, viscous secretions (1). The hallmark signs and symptoms of cystic fibrosis are salty tasting skin, poor growth and poor weight gain despite a normal food intake, accumulation of thick, sticky mucus, frequent chest infections, and coughing or shortness of breath. Males can be infertile due to congenital obstruction of vas deferens. Symptoms often appear in infancy and childhood, such as bowel obstruction due to meconium ileus in newborn babies (2, 3). Cystic fibrosis may be diagnosed by many different methods including newborn screening, sweat testing, and genetic testing (4). The prognosis for cystic fibrosis has improved due to earlier diagnosis through screening, better treatment and access to health care (5). This disease is specified by very wide spectrum of clinical manifestations for which many differential diagnoses have to be considered. We studied 300 patients of CF in Children's Medical Center in Tehran, during 2006 to 2012. Necessary information including age, sex, and symptoms of patients such as cough, hemoptysis, recurrent pulmonary infection, nasal polyposis, recurrent sinusitis, intestinal obstruction, intussusception, reflux, rectal prolapse, pancreatitis, fatty diarrhea, edema and hypo-albuminemia, anemia, vomiting and dehydration attack, cirrhosis, biliary atresia, portal hypertension, salty skin, failure to thrive (FTT), clubbing, coagulation disorder, bone fracture and nyctalopia was entered in the checklists. First manifestation of each patient was also noted. The main complaint of patients was isolated FTT and this complication had occurred in 74 (24.7%) patients. In addition, FTT with another complication was observed in 28 (9.3%) patients at the beginning of the disease; in other words, isolated failure to thrive or FTT associated with other symptoms of the disease was seen as first clinical presentation in about one-third of the patients (that is, 102 patients). The most common complication next to FTT was pulmonary infection which was seen in 47 (15.7%) patients. Other common first clinical presentations of cystic fibrosis include: attacks of vomiting and dehydration in 45 (15%), cough in 25 (8.3%), intestinal obstruction (meconium ileus) in 23 (7.7%), fatty diarrhea in 18 (6%), FTT with fatty diarrhea in 11 (3.7%), edema and Hypo-albombinemia in 10 (3.3%), and biliary atresia in 6 (2%) patients. Other symptoms including anemia, salty skin, coagulation disorders, gastro esophageal reflux, and sinusitis were seen in less than five patients. In 30 (10%) patients, two of the mentioned complications were observed simultaneously. At onset of disease, among a total of 300 patients, 182 (60.7%) had gastrointestinal symptoms, and 72 (24%) patients had respiratory tract symptoms. CF may be diagnosed after a long period of illness, by different symptoms and laboratory tests (6). In a similar study by Fallahi et al. in Iran, prevalence of first clinical presentations of CF patients before the diagnosis was as follow: gastro intestinal symptoms in 152 (62.6%), respiratory symptoms in 93 (38.3%), isolated FTT or in combination with other symptoms in 34.9%, diarrhea in 28.8%, vomiting in 15.6%, cough in 30% and respiratory distress in 23.9% patients (6). As mentioned before, the first complication in 7.7% of our patients was ileus meconium which is similar to results of other studies (4). According to this study, gastrointestinal symptoms were the most common symptoms associated with CF disease. In a study by McCloskey more than two thirds of patients presented with respiratory symptoms including chronic sino-pulmonary disease, nasal polyps or recurrent respiratory tract morbidity. This study tried to bring up the

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most common presenting symptoms of CF. With more attention to the symptoms of this disease, the diagnosis could be established earlier and upcoming complications prevented.

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