

Letter to the editor

Cystic fibrosis as a mimicker of inborn errors of immunity: the importance of considering mental health

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Dear Editor

Inborn errors of immunity (IEI) are characterized by a weakened immune response leading to frequent repeated infections (e.g. pneumonias, sinusitis, bronchitis, ear infections, meningitis, and skin infections).¹ Because of such a wide spectrum of manifestations, a variety of differential diagnoses must be kept in mind at dealing with these children. One of them is cystic fibrosis.

Cystic fibrosis (CF) is an autosomal recessive genetic disorder that leads to increased susceptibility to recurrent pulmonary bacterial infections.² Beside the lungs, the pancreas, liver, kidneys, and intestine are commonly involved in such a chronic disease.³ The usual onset of symptoms is during the first 6 months of life, but it could be delayed to adolescence or adulthood in milder forms.⁴

Thick mucus accumulation is a pathognomonic feature of CF affecting different organs especially the lungs with subsequent dyspnea and productive cough. On the other hand, sinusitis, impaired growth, steatorrhea, male infertility/low female fertility, and clubbing of fingers and toes are commonly encountered in affected patients with different frequencies and severities according to the type of the underlying gene mutation.^{2,4} These chronic manifestations make it a mimicker of some IEI.

CF manifestations are exhausting and debilitating representing a very stressful lifelong condition leading to increased vulnerability to internalizing mental disorders but because of the recent improvement of the availability and efficacy of health care services provided to patients, survival rates of such cases increased.⁵ Consequently, mental health and health related quality of life have become a crucial issue in their management. Proper psychometric assessment and implementation of psychotherapy can improve their quality of life and empower them to be independent, productive, and optimistic.^{6,7} With such progress in mental management, CF patients started to see the light at the end of the tunnel.

Zaky et al.⁷ studied 31 clinically and laboratory verified Egyptian children and adolescents with cystic fibrosis compared to age and sex matched healthy controls using Children Depression Inventory Scale, Spence Children Anxiety Scale, and the Pediatric Quality of Life Scale. They found significantly higher frequency of depression and anxiety among cases compared to controls explained by the stress of their chronic disease and the burden of being compliant to its management. On the other hand, there were insignificant differences between studied groups regarding total score of the Pediatric Quality of Life Scale and its subscales that was indicative of the efficiency of the implemented CF therapeutic plan in controlling their symptoms and improving their life quality.

In conclusion, pediatric immunologists need to be aware of the overlap of clinical manifestations between IEI and CF and the necessity of including psychometric assessment and psychotherapy as well as the intake of psychopharmacological medications whenever indicated as an integral part of their multidisciplinary management plan. Always remember, whenever and wherever there is help, there is hope.

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