BÖLÜM 3

ÇOCUKLARDA KİSTİK FİBROZİS İLİŞKİLİ DİYABET

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GİRİŞ

Kistik fibrozis (KF), 1:2500-5,000 canlı doğum prevalansı ile Hispanik olmayan Beyaz ırkta en sık görülen ölümcül otozomal resesif bozukluktur (1). KF, Kistik Fibroz Trans- Membran Regülatörü (KFTR) genindeki mutasyonlardan kaynaklanır ve çoklu organ tutulumu ile karakterizedir. Etkilenen bireylerde klinik belirtilerin şiddeti, altta yatan genetik kusura bağlı olarak değişir. Başta erken tanı, hiperkalorik diyet ve alevlenmelerin agresif tedavisi olmak üzere birçok faktör, hastaların medyan yaşam beklentilerinde bir kaymaya yol açmıştır (2). Yaşam süresinin uzaması, kistik fibrozis ilişkili diyabet (KFRD) prevalansının artmasına neden olmuştur (1, 3). Glikoz metabolizmasındaki bozukluklar daha genç yaşlarda da ortaya çıkabilir (çocukların %2'sinde), ancak hastaların çoğuna ergenlik döneminde veya yetişkinlikte (tüm vakaların %40'ına kadar) KFRD tanısı konur (4). KF hastalarında glukoz homeostaz bozukluklarının tanınması, tedavisinde doktorların yaklaşımı ile ilgili çalışma ilk kez 1998 yılında yapılmış, bozulmuş glukoz toleransı ve diyabetin taranması ve tedavisindeki farklılıkların kaldırılması için bir konsensus sağlanması gerektiği bildirilmiş, sonraki yıl KFRD'nin tanı ve tedavisi için ilk klavuz yayınlanmıştır (5,6).

EPİDEMİYOLOJİ

KF hastalarında beklenen yaşam sürelerinin artması ve KFRD farkındalığının, yapılan taramaların artması ile komplikasyonun görülme sıklığında da artma

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