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Original Article

Long-term follow-up of cystic fibrosis newborn screening: Psychosocial functioning of adolescents and young adults

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Abstract

Background: Long-term psychosocial outcomes of cystic fibrosis (CF) patients diagnosed through newborn screening remain unknown.
Methods: This cross-sectional study compared three groups of youths (16 to 22 years): CF patients diagnosed through NBS (CF-NBS, n = 13), CF patients diagnosed through standard practice (CF-SP, n = 26) and healthy peers (H, n = 42), plus 72 of their parents. We hypothesized that adolescent psychological functioning would be mediated by parent depression and quality of parent–child communication and cohesiveness.
Results: A path analysis showed significantly more depression among CF-NBS group parents (p = .006–.008). Parent–child cohesiveness was related to communication (p < .001). Cohesiveness and communication were associated with youth Internalizing Problems (p = .037, p = .009), Emotional Symptoms (p = 0.018, p = 0.022), and Personal Adjustment (communication only, p = 0.009). Parent depression was related to youth Personal Adjustment (p = 0.022).
Conclusions: CF patients report psychosocial function similar to healthy peers. Parents of children diagnosed with CF through NBS may be at risk for depressive symptoms when their children reach adolescence.

Keywords: Adolescence; Cohesiveness; Communication; Cystic fibrosis; Newborn screening; Psychosocial function

1. Introduction

Early diagnosis of cystic fibrosis (CF) resulting from newborn screening and innovative treatments have led to increased longevity for people with CF. Affected families also face new challenges as well as opportunities as a consequence of such advances. Mothers of newborns with CF tend to choose bottle feeding over breast feeding [1]. Parents of young children with CF reportedly perceived their children as being more vulnerable to illness than parents whose infants either have no health problems or have another health conditions identified through newborn screening, e.g. congenital hypothyroidism [2]. Evidence also shows that a CF diagnosis made during early infancy can be associated with parental depressive symptoms [3,4]. Maternal depression can adversely affect mothers' capacities to provide their infants with sensitive and responsive caregiving [5], which provides the foundation for infants to form secure attachments with their parents and close interpersonal relationships later in life

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Abbreviations: CF, cystic fibrosis; NBS, newborn screening; ADHD, attention deficit hyperactivity; RCT, randomized control trial; IRB, Institutional Review Board; BASC-2, Behavioral Assessment System for Children — Second Edition; PRQ, Parenting Relationship Questionnaire; MCAR, missing completely at random

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[6]. Toddlers diagnosed with CF in early infancy (due to the presence of CF symptoms) were observed to have higher rates of insecure infant–mother attachments than children diagnosed with CF later in infancy [7]. These findings raise questions about the long-term impact of a neonatal CF diagnosis resulting from NBS on the quality of parent–child relationships and youth psychosocial functioning later in life.

1.1. Psychological functioning of adolescents with CF

As children with CF reach adolescence they often experience disease progression that forces them to rely more heavily on their parents for health care during a developmental phase that typically involves striving for individuation from parents [8,9]. Concurrently, they should be shifting their attention to strengthening peer relationships. However, school absence due to illness and hospitalization limits access to friends [10], while medical protocols, designed to prevent cross-infection, preclude CF patients from socializing with one another. Female teens, who tend to have more severe lung disease than males [11], may be particularly susceptible to stressors associated with self-care and social isolation [12]. Not surprising, children and adolescents with CF often report higher rates of depression and anxiety than the general population [13] which is also associated with non-adherence to airway clearance and insecure attachment to parents [14]. One study reported 32% of adolescents and young adults with CF had clinical levels of anxiety and 3% had clinical levels of depression [15]. Another study showed anxiety and depression in adolescents and young adults with CF to be lower than in healthy controls [16]. One report showed 10% of CF patients ages 5–18 years met the diagnostic criteria for attention-deficit hyperactivity, 60% of whom were non-adherent to prescribed CF treatment [17]. This rate of ADHD in CF patients was higher than the 6–7% reported in the general population [18]. Studies of youths with ADHD not associated with CF suggested that the quality of parent–child relationships can affect the symptom manifestation and social functioning of individuals with ADHD [19]. While adolescents with CF may be at risk for psychosocial problems, the mechanism for developing such symptoms remains unclear.

1.2. Parent psychological functioning and family relationships

Parents of children with CF have reported more overall stress and more illness-related stress than parents of healthy children [20,21]. Mothers report anxiety and depression; fathers report depression and other Internalizing Problems [21–23]. It has been suggested that parental anxiety and/or depression can lead to similar symptoms in their children [24].

Family cohesion, expressiveness, and organization, as reported by children with CF, have been negatively correlated with child self-reported anxiety and depression [16]. Family cohesiveness, flexibility, and positive interactions have been associated with adherence to treatment as reported by children with CF and their parents [25,26]. The quality of parent–child relationships represents a potential mediating factor in the psychological well-being of youths with CF.

The quality of parent–adolescent relationships in normative samples has been found to affect various aspects of teens' psychosocial development [27,28]. It is well documented that secure attachments (particularly to mothers) allow adolescents to move towards cognitive and emotional autonomy [27,29]. Adolescent attachment security has been correlated with the capacity to effectively communicate emotion [30]. Insecure parent–child attachment has been linked to adolescent internalizing symptoms [28] and discrepancies between adolescent self-report and parent report of adolescent psychological symptoms [31]. Thus, research with normative samples and patients with CF points to the quality of parent–child relationships as a critical factor influencing adolescent psychosocial functioning.

This study drew from the Circumplex conceptual model of family systems [32] to examine the relationship between a neonatal CF diagnosis resulting from NBS and youth psychosocial functioning in late adolescence and early adulthood. Two central concepts in this model are communication and cohesiveness. Communication includes sharing thoughts and feelings, respectful listening, and empathy for other's feelings and experiences. Cohesiveness is the affective bond or emotional attachment between family members. Effective communication facilitates cohesiveness. We posit that a neonatal diagnosis (from NBS) is likely to produce parental distress and perceptions of child vulnerability that could adversely affect the quality of parent–child cohesiveness and patterns of communication early in the child's life and continue well into the child's adolescence. Given the progressive nature of CF, parents are likely to experience either on-going or recurrent emotional distress. Observing their children's repeated pulmonary infections and/or emergence of new CF complications is likely to reinforce parents' perceptions of their children's vulnerability. Consequently, patterns of interaction between parent and their children that transpire from a neonatal diagnosis are likely to become entrenched in ways that could affect children's long-term psychosocial development. We hypothesized that adolescent psychosocial functioning would be mediated by parent depressive symptoms, parent–child communication, and parent–child cohesiveness. We also controlled for child age and gender, and parent education based on previous research.

2. Methods

This cross-sectional study was conducted within the Wisconsin NBS Project, a longitudinal evaluation of benefits and risks of NBS for CF [33]. The original study, conducted from 1985 to 1994, involved random assignment of newborns into two groups: infants diagnosed through NBS and infants diagnosed through methods that were standard practice at the time, e.g., symptom development and/or family history. Median age at diagnosis was 6.9 weeks (range = 3 days to 5.4 years, including 5 false-negatives) for the NBS group and 24 weeks (range = 4 days to 15.7 years) for the standard practice group. Parents of 145 children with CF, confirmed by sweat chloride levels of ≥ 60 mmol/L, consented