

Psychological Impact of Eosinophilic Esophagitis on Children and Families

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KEYWORDS

- Eosinophilic esophagitis (EoE) • Children and adolescents
- Chronic illness • Psychological adjustment • Quality of life

Eosinophilic esophagitis (EoE) is a newly recognized disorder marked clinically by symptoms of upper gastrointestinal distress and by histologic findings of increased eosinophils in the esophagus.¹ Recent reports have documented that EoE occurs in children and adults, with initial diagnoses made in children as young as 6 months of age.¹ Most EoE symptoms among children are triggered by allergic hypersensitivity to foods.² For pediatric patients and their families, symptom experiences, diagnostic procedures, and recommended treatments related to the disease can have a tremendous impact on quality of life and on psychological and social adjustment. However, the manner in which parents and children respond to, and cope with, elements of the disease can significantly influence the impact of EoE on resulting quality of life. The goal of this article is to describe how the EoE disease experience affects the quality of life of children and their families, and to raise key questions regarding the unique physical and emotional challenges posed by this disease.

Because EoE in children has only recently been recognized and defined as a clinical syndrome, much about the natural history of the disease has yet to be discovered. Similarly, although some clinical studies have investigated treatment strategies for differing clinical presentations, much remains to be learned about optimal treatment. Systematic research is lacking regarding the psychological effects of EoE on children and the impact of the disease on children's quality of life and that of their families. However, some lessons can be learned by drawing on existing research regarding health-related quality of life (HRQL) for children who have similar chronic illnesses. By combining existing research findings with the clinical experience of a pediatric behavioral health team that has worked with children with EoE and their families, it is

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possible to formulate a view of the impact of EoE on quality of life and the primary coping challenges faced by these children and their families.

Research regarding other pediatric chronic illnesses has shown that the disease experience depends on multiple factors. Studies of HRQL among children who have medical illnesses are concerned with the effects of the disease and associated medical treatments not only on physical status, but also on the psychological and social aspects of children's lives.³ Chronic illness influences children's psychological adjustment, with short-term effects on emotional responses to symptoms and treatment, and long-term effects on overall adjustment.⁴ Although the presence of a chronic illness has several general effects on children and families, other effects of pediatric chronic illnesses are related to the unique characteristics of the disease and the treatments.⁴ Effects also depend on the developmental status of the affected child and on the life context being considered, such as the family environment within which children and adolescents experience their disease. Finally, chronic illness in children also has major effects on the children's caregivers.⁵ Thus, it can be expected that the experience of having EoE will have effects on quality of life similar to those of other diseases, along with effects that are unique to EoE; that those effects will differ, depending on the characteristics of individual children and their families; and that caregivers will be greatly affected, in addition to the children and adolescents diagnosed with EoE.

QUALITY-OF-LIFE EFFECTS OF EOSINOPHILIC ESOPHAGITIS SYMPTOMS

In infants and young children, EoE most commonly presents as reflux or vomiting, early satiety, feeding aversion or intolerance, and failure to thrive.¹ Many of these symptoms are indistinguishable from those of gastroesophageal reflux disease (GERD); only the presence of increased numbers of eosinophils and the failure to respond to standard treatment of GERD indicate that the underlying disease is EoE.¹ As with GERD, symptoms in infants and young children are observed and reported by caregivers. When infants spit up or vomit, or they are fussy and irritable, their caregivers infer that they are experiencing discomfort or pain. Food aversion or intolerance are believed to stem from experiences of discomfort or pain with eating, leading to learned avoidance behavior in children too young to verbalize their pain. In fact, infants who have GERD during their first year of life were significantly more likely than healthy children to develop feeding problems within the following year,⁶ and it is likely that infants who have EoE have a similar pattern. Because of infants' and toddlers' developmental status, their EoE symptoms, including reflux, food aversion, and failure to thrive, have a major impact on the quality of life of their caregivers. The impact on caregivers' quality of life will be discussed later, when the effects of EoE on families are addressed.

In children who are of school age and older, EoE symptom patterns are different from those of younger children, which, in part, may be because of the older children's ability to report internal sensations, but also because the disease may manifest differently with increased age.¹ Typical symptoms among older children and adolescents include heartburn, abdominal pain, early satiety, reflux, and vomiting. In addition, older children experience dysphasia (difficulty swallowing), choking, or esophageal food impaction. As with younger children, a large overlap with GERD symptoms occurs; however, those symptoms that are different from typical GERD symptoms and are most often associated with EoE, such as difficulty swallowing and food impaction, can be extremely distressing and may have significant effects on psychological functioning in older children and adolescents.

Research information is available regarding quality of life for children who have several gastrointestinal diseases, including GERD, and may be applicable to those children whose EoE symptoms mimic those of GERD. Using a generic instrument, children aged 5 to 18 reported on their quality of life in the domains of physical, emotional, social, and school functioning.⁷ Their quality of life scores were comparable to those of children and adolescents who have diagnoses of inflammatory bowel disease and functional abdominal pain, and all were somewhat lower than scores for a healthy control group. Although these results confirm that children who have GERD symptoms experience a decrement in quality of life similar to that of other gastrointestinal conditions, they nevertheless provide limited insight into quality-of-life effects that are specific either for children who have GERD or for those who have EoE with GERD-like symptoms. To learn about the impact of symptoms on quality of life among children who have EoE, it is necessary to learn which children are experiencing specific EoE symptoms, the symptom frequency and severity, and the associated emotional distress and social and psychological impact.

Swallowing problems have been commonly reported among older children and adolescents who have EoE. The severity of swallowing problems varies a great deal, as does the children's related distress. For example, an 11-year-old boy said that having EoE seldom bothered him. About once a month he felt as if a ball were in his esophagus but he was able to eliminate the sensation by taking a sip of water. In contrast, another 12-year-old boy, in describing his "throat problem," had reported difficulties swallowing and choking on a daily basis. As a result, he never ate solid food at school, instead drinking only milk. His school friends were aware of his tendency to choke and looked out for him. At home, he ate small amounts of food continuously because he was often ravenously hungry. For the latter patient, the swallowing problems that characterized his EoE had a great impact on his day-to-day life in terms of his eating behavior and his nutritional intake; however, his emotional response to the symptoms appeared matter-of-fact. For other youngsters, the experience of choking on food on a regular basis is more than unpleasant; it is extremely frightening. It is not uncommon for children and adolescents who have these symptoms to develop strong anxiety reactions related to swallowing food. The experience of having a food impaction may be especially traumatic. For example, following an impaction and the emergency procedure required for removal of the food from his throat, an older adolescent began having panic attacks, and he required psychological treatment to help him eat without anxiety.

In sum, available information suggests that EoE symptoms affect children and adolescents' quality of life, and also possibly their psychological adjustment. However, no systematic information is available about the frequency with which various EoE symptoms are associated with meaningful effects within the key domains of child functioning across developmental stages. It appears that persistent symptoms in infancy may put children at high risk for feeding disorders, but the frequency and persistence of this outcome is unknown. It is unknown what level of swallowing difficulty or regular choking affects a child's or adolescent's nutritional status or social relationships. Similarly, no information is available regarding the emotional sequelae of an episode of food impaction, such as the frequency with which anxiety reactions occur. It would be helpful to discover the coping strategies that children and adolescents use when they experience symptoms, and their effectiveness in managing them. Thus, many questions remain regarding the relationship between the symptoms experienced by children who have EoE and the effects on quality of life and psychological adjustment.

QUALITY-OF-LIFE EFFECTS OF EOSINOPHILIC ESOPHAGITIS TREATMENTS

Children and adolescents who have EoE are impacted not only by the disease symptoms but also by the treatment of the disease. Recommended treatments of EoE in children usually involve medication or dietary manipulations.¹ Pharmacologic treatment may include reflux medications or swallowed topical corticosteroids. Because the removal of causative foods from the diet has been clearly demonstrated to be successful in the treatment of EoE, dietary elimination is a key component of most children's experience with EoE. Dietary interventions range from the elimination of several specific foods from the diet, to removal of about six of the most common allergenic foods, to allowing ingestion of only elemental formulas. The time course of dietary restrictions can range from a few days (eg, for trials of specific foods) to weeks, months, or even years. To provide sufficient calories with the elemental formulas, it is common to place a nasogastric tube for feeding for a limited time. When an elemental formula diet is deemed necessary over a longer period of time, a gastric tube (G-tube) may be placed directly into the stomach. Thus, treatment strategies vary in their bodily invasiveness and in the extent of required lifestyle changes.

The impact on children and adolescents of diagnostic procedures and treatments of EoE varies widely. In general, the ways in which children respond to procedures such as blood draws, skin testing, and endoscopies and to swallowed medications related to EoE appear to be similar to the ways in which children cope with procedures and treatments of other diseases.⁸ The psychological impact depends on the individual child's age and stage of development, especially on the ability to understand what is involved in the procedure or treatment, and the reasons for it. Much also depends on the quality of emotional communications and coping assistance provided for the child by parents. Apart from parenting methods, children's temperamental styles are a key determinant of their response to procedures and treatments. A child who tends to be emotionally reactive, anxious, or inhibited responds differently from one who is flexible and easy-going, and a robust and strong-willed child will have yet a different response.

Although many of the EoE procedures and treatments, such as diagnostic tests and medication administration, might be considered routine, the illness-specific dietary restrictions that comprise a primary treatment strategy for children and adolescents who have EoE frequently pose a unique and difficult psychological challenge for children and their families. Extensive restrictions on children's diets may have the potential for a severe negative impact on all components of HRQL for children: their physical, emotional, social, and school functioning. Several aspects of dietary restrictions make this treatment particularly difficult. Eating habits and favorite foods are pervasive and deeply emotional aspects of children's lives. Family routines and social events alike revolve around meals and treats. Given the centrality of food in children and families' lives, dietary restrictions have a major impact on children who have EoE in much the same way as for children who have food allergies. However, for children who have EoE, a wide range of foods is often eliminated, in comparison with one or several foods for food allergic children. Also, the consequences of eating the offending foods are often not readily obvious to children who have EoE in the way that immediate reactions are to children who have food allergies so, unless they experience dramatic symptoms such as vomiting or food impactions, children who have EoE may fail to make the connection between their dietary restrictions and fluctuations in their disease. Given these factors, elimination diets that are central to the treatment of EoE may have a profound effect on children's quality of life.

Not every child responds negatively to implementation of an elimination diet. Clinical experience has shown that infants and very young children who require an elemental formula often do well. Their mothers point out that they have never known anything different, and they are enormously relieved when the symptoms remit and the child begins to thrive. The elimination of foods can be more difficult for toddlers who have begun eating solids, who are often selective in their food choices, and for whom withdrawal of a favorite food can lead to distress. For them, the experience of having new foods introduced and then removed from their diet, a process that occurs with food trials, can be particularly difficult, and reducing the emotional impact requires a great deal of parental and professional strategizing.

For preschool-aged children, the implementation of a restricted diet is often extremely difficult. At this age, children's understanding of their bodies is expanding, their emerging sense of self is facilitated by the discovery of favorite foods, and development of the ability to regulate emotions and behavior is intertwined with their expectation and mastery of routine. Although cognitive abilities are expanding, preschoolers are as yet unable to understand the rationale for the imposition of dietary restrictions. For example, a 4-year-old girl was accustomed to a wide range of food, but her frequent vomiting dictated the elimination of dairy and wheat from her diet. Her response was pervasive, with extreme anger at friends and family members, especially at her younger 2-year-old brother who was allowed to eat what he pleased. The brother, on the other hand, stopped eating what he was allowed and began restricting his diet to match that of his sister (illustrating one type of effect elimination diets can have on siblings!).

Despite their greater cognitive abilities, older children also can have much difficulty when food is partially or completely removed from their diet. Children may become anxious and depressed, or they may become oppositional. One child stole food from other family members, and her parents locked their kitchen cabinets to keep her from eating prohibited food. On the other hand, it is not unusual for teenagers, who typically have independent access to food, to angrily disregard the dietary prohibitions to be able to eat favorite foods such as pizza.

Some children who have EoE require gastrostomy tubes (G-tubes) to ensure that they receive adequate nutrition. Little information is available regarding the quality-of-life effects of tube feeding for patients whose lives are not in immediate danger. It may be that psychological adjustment in children who have EoE and have G-tubes is dependent on the same factors that affect children's adjustment to other treatments. These include the child's age at the time the G-tube is placed, the child's temperament and psychological adjustment before G-tube placement, and the family emotional context. For example, a 2-year-old vomited so frequently that he failed to gain weight over a 10-month period, and his cognitive and emotional development was compromised. His mother was vastly relieved when a G-tube was placed. Along with weight gain, this child evidenced brightened affect and accelerated cognitive and motor development. Unfortunately, the reinstatement of oral feeding can be extremely challenging for children, because of either recurring symptoms or learned patterns of food refusal.⁹ Many questions remain regarding the short- and long-term emotional and social consequences of the use of G-tubes in the treatment of children who have EoE.

For children and adolescents who have EoE, quality of life is further impacted by additional medical disorders. Seventy percent of children diagnosed with EoE have one or more coexisting atopic disorders such as asthma, atopic dermatitis, allergic rhinitis, or food allergies.^{1,10} Each of these disorders presents with symptoms that require attention and a treatment regimen, so that the coexistence of EoE with

additional atopic disorders makes for a highly complex disease experience, impacting quality of life in multiple ways. The coexistence of EoE and food allergy can be a particularly challenging combination. Children who have EoE and immediate hypersensitivity food allergies are restricted from a wide array of foods, with varying levels of negative effects, including the possibility of anaphylaxis. An anxious 8-year-old boy who had both conditions explained: "Some food makes me sick if I eat it; some other food, if I eat it I could die." Clinical observations suggest that the combination of EoE and food allergies increases the likelihood of symptom confusion and high anxiety related to food exposure.

Children's characteristic temperamental style before onset of EoE is an additional influence on their response to the disease. From infancy onward, children exhibit distinct tendencies to respond to their environment in characteristic ways. Infants may show behavioral inhibition, or wariness, that often is manifested as shyness, or even anxiety, as they get older. Young children may respond to circumstances in an easygoing, flexible manner, or they may tend to be more emotionally reactive, sensitive, or inflexible, requiring much parental modulation to navigate their daily lives. With the onset of EoE, children's predisposing temperamental characteristics influence their experiences of and responses to their internal sensations such as those of hunger or pain, to diagnostic procedures such as skin testing or endoscopies, and to the changes in their routines necessitated by treatments such as elimination diets. As children get older, those who have tendencies to anxiety or depression have increased vulnerability when they are faced with elimination diets or invasive medical procedures. For one 9-year-old boy who had EoE triggered by hypersensitivity to almost all foods, and who also had food allergies, treatment consisted of eliminating all orally ingested foods and placing a G-tube to meet his nutritional needs. Unable to attend school or engage in activities with peers, this emotionally reactive and anxious child became profoundly depressed. His depression subsided somewhat when orally ingestible food was reintroduced into his diet, but his anxiety level remained high and he required further treatment to address his emotional response. In this manner, children and adolescents who are already vulnerable to stress because of their tendency toward anxiety or depression may develop serious psychological reactions when faced with deprivation or trauma; others may be able to take such events in stride.

To summarize, the treatments of EoE can have a significant impact on children's quality of life, although the effects depend on both child and family factors. Because of the centrality of food in children's lives, elimination diets have an especially great potential to profoundly impact children's adjustment. Research is needed if we are to better understand the psychological effects on children of elimination diets and to weigh them against the improvement in quality of life brought about by short-term symptom reduction and the prevention of long-term physical effects on the gastrointestinal system.

THE DISEASE EXPERIENCE FOR FAMILIES OF CHILDREN WHO HAVE EOSINOPHILIC ESOPHAGITIS

Chronic illness in children impacts not only the quality of life of the ill child but also that of the parents and siblings. Parents of chronically ill children report increased emotional strain and financial burdens, as well as effects on relationships within the family and social contacts outside the family.⁵ In addition to providing information on children's quality of life, studies of similar diseases, such as GERD and food allergy, give insight into the effects on parents and siblings of children who have EoE.

In a study of caregivers of young children who had GERD, investigators conducted focus groups to identify issues affecting the parents' quality of life.¹¹ Parents talked about how routine childcare activities for children who have GERD required extra work and care, such as special feeding techniques involving small portions, frequent feedings, and preparation of special meals or formulas. They needed to spend additional time making meals because of their children's dietary needs, and they had extra housework because of soiled clothing or furnishings. The parents also talked about emotional concerns specific to their children's reflux and feeding problems. For example, they expressed fears that the child might choke on their vomit, especially when they put them to bed at night. Parents of children who had food refusal and failure to thrive, in particular, worried a great deal about their children's condition and prognosis. This finding is consistent with other studies that have documented high levels of parenting stress among parents of children who have feeding disorders.¹² Although feeding tubes can initially bring relief to parents concerned about their children's nutritional intake, one study showed that parents of children who had G-tubes had poorer quality of life in terms of social life, family life, sex life, and work.¹³ The additional burden and high levels of emotional stress reported in these studies are consistent with clinical experience with parents of children who have EoE.

Because treatment of EoE typically involves dietary restrictions, many of the lifestyle changes required for families of children who have EoE are similar to those faced by families of children who have food allergies. However, although parents of food-allergic children often suffer from anxiety related to the possibility of life-threatening reactions, central worries for parents of children who have EoE concern avoiding precipitating foods while meeting their children's nutritional needs. A study of the impact of food allergies on families' lives investigated the effects on daily care giving and on family members' emotional lives.¹⁴ As expected, meal preparation and family social activities were most strongly affected. However, caregiver stress received high impact ratings, and a sizable proportion of the caregivers reported large effects on sibling and spousal relationships. The diagnosis and management of EoE appear to be accompanied by significant increases in caregiver burden, parental anxiety and distress, and strain and tension within couples' relationships.

Caregiver anxiety and relationship strain are worsened by uncertainty related to children's chronic illness.⁵ Because EoE is newly recognized, limited information is available about the natural history of the disease or about the short- and long-term effects of treatment, leaving parents in an even greater state of uncertainty than that experienced by parents of children who have well-characterized pediatric illnesses. The effect of the uncertainty surrounding EoE becomes apparent when compared with families' experiences with better known diseases such as diabetes, where diagnostic tests are available, risk factors and prognosis are known, treatment has been agreed on, educational and coping programs are advanced, and even psychological contributions and ramifications are known. In contrast, diagnostic criteria for EoE have only just been determined, and because the condition is less well known among general practitioners, parents have found themselves searching the Internet for an explanation for their children's symptoms. Parents report experiencing considerable stress during the process of trying to decipher their child's medical problem (eg, when reflux symptoms persisted despite routine treatment). Once a diagnosis was made, parents became aware that experts could disagree about optimal treatment, engendering further uncertainty and anxiety.

Despite limited information available about EoE, much variability exists in levels of distress and anxiety that parents experience in response to their children's disease. This variability may be related, in part, to differences among individuals in their

tolerance for uncertainty. In general, high levels of uncertainty intolerance are related to anxiety, worry, and depression, and a need for cognitive closure.¹⁵ For such parents, the current dearth of available information is intolerable, and they cope by searching for any available information, whether it portends positive or negative outcomes. For example, because little is known about the natural history of EoE, parents' natural inclination to prepare for their children's future motivates them to seek information from other parents whose children may have a much different disease profile. Having some information, even if it erroneously indicates a negative outcome, allows individuals who have little tolerance for uncertainty to experience anxiety-relieving cognitive closure. Strain in marital relationships may be exacerbated by parents' anxiety, especially when spouses' use of antithetical coping strategies leads to conflict. For one couple, conflict ensued when the mother sought information on the Internet regarding prognosis and treatment options, whereas her husband questioned the validity of the EoE diagnosis and the necessity of treatment beyond symptom management. An intervention aimed at increasing uncertainty tolerance and reducing anxiety may provide help for some anxious parents of children who have EoE to cope more effectively.

To summarize, EoE may have a profound effect on the quality of life and psychosocial adjustment of parents and siblings of the affected child. Much time and work is required to decipher and monitor symptoms, to meet dietary requirements, and to plan medical follow-up, resulting in tremendous emotional and financial stress. The emotional stress and resultant strain on family relationships is exacerbated by the limited information currently available about the natural history and treatment outcome of EoE in children and adolescents.

SUMMARY

Because EoE has only recently been recognized and described, systematic research regarding the natural history of the disease and the short- and long-term effects of treatment is in its infancy. Although no research has addressed the psychosocial impact of EoE, clinical experience indicates that disease symptoms and treatments can have profound effects on the quality of life of affected children and their families. The responses of children and adolescents are variable, and are dependent on developmental level, temperament, and pre-existing psychological adjustment. Although parents of chronically ill children typically experience increased burden and stress, it is possible that the uncertainties currently associated with EoE contribute to even higher levels of anxiety. Research studies are needed to investigate the impact of EoE symptoms and of current treatments on quality of life and psychological adjustment in children and their families, and to discover how best to help families cope in an optimal manner.

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