



# Complex Child E-Magazine

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## Living with Eosinophilic Esophagitis by Darshani Sukumaran



The author (second from left), with her daughters Nitara (left) and Abirami (center), her husband Vignesh, and a few cuddly companions

Last summer I found myself in Utah at a special conference. Surrounded by children and adults with feeding tubes attached to small whirring backpack pumps, I felt strangely at home. I never thought that I would feel at home amongst people with feeding tubes! Welcome to my new normal. This conference, sponsored by APFED (American Partnership for Eosinophilic Disorders), brought together people who suffer from a family of conditions called Eosinophilic Gastrointestinal Diseases. The most common manifestation is Eosinophilic Esophagitis, or EE. The journey to Utah was for me a type of homecoming. For the first time since I had my daughter Nitara I felt that I was in the company of people who knew exactly what we had gone through with her.

EE is a disease that was discovered just twenty years ago, thanks to modern technology and improved techniques of diagnosis. The eosinophil is a type of white blood cell that is

made in bone marrow. Mature eosinophils migrate to the intestines where they reside in small numbers, waiting to attack parasites. In people with EE something goes wrong: the eosinophils attack foods instead.<sup>1</sup> The proteins in foods trigger a localized immune response, causing colonies of eosinophils to flock to the esophagus. The continued ingestion of food exacerbates the immune response and soon the tissues become swollen and damaged. Over time, this leads to permanent scarring and a loss of tissue flexibility. A person with prolonged EE can develop rings, furrows, a thickened esophagus, and impaired swallowing function (dysphagia).<sup>2</sup>

Because EE mimics Gastroesophageal Reflux Disease (GERD), EE can go undiagnosed for years. EE is making headlines in recent medical journals, but it is still a relatively unknown disorder to primary care providers and even specialists. EE requires biopsies to be taken from specific areas of the GI tract and examined by a pathologist who is familiar with the condition. A normal count of eosinophils in the esophagus is zero to four. A person with severe GERD may have up to 10. If the count is greater than 15-20 then a diagnosis of EE is given.<sup>3</sup> My daughter Nitara had over 100 eosinophils on the biopsy that diagnosed her EE.

When my daughter was born, she showed an immediate aversion to feeding. By a few days old she was clearly in discomfort and none of the usual comfort measures helped. I saw her transform from a healthy baby to one who was very thin and withdrawn. After repeated visits to the pediatrician, who told me it was just colic, I finally took her to the ER one Saturday night. She had not fed for most of the day and screamed non-stop. At three months old she was only taking 12-15 oz of formula on a good day. Nitara was immediately admitted with dehydration and failure to thrive. The GI on call said she was exhibiting Sandifer's Syndrome or classic reflux positioning, common for refluxers. Six days later, Nitara came home with a feeding tube down her nose (NG tube) and prescriptions for reflux medication.



Nitara as a baby after getting her NG tube

She was finally gaining weight and seemed a bit happier on the medication. Unfortunately, the vomiting continued to be a constant in her life. At eight months old she had a permanent G-tube inserted into her stomach. Two biopsies showed GERD but

not EE. It either had not developed yet or the pathologist did not find it. Nitara had a major feeding aversion that was helped by therapy somewhat. She was very brave to continue trying to eat even though it hurt her and made her gag. Even though she was gaining weight, Nitara was not able to fight infections as well as she should have. She was plagued with ear infections, repeated colds, high fevers, febrile seizures, croup, and of course vomiting that worsened any time she was sick. She was diagnosed with asthma after getting RSV at age two. Finally, at 35 months I took her to her third GI, one who I later found out cares for many EE patients, and she was diagnosed with EE. Nitara still has classic GERD even when the EE is in remission, which is probably the reason her previous doctors did not think to look for the EE.

EE shows different symptoms at different ages. Throat and intestinal pain is present in all ages. Babies show difficulty or reluctance to feed.<sup>4</sup> By their first birthdays they vomit excessively, have stomach pain, feeding aversions, and if the eosinophils are further down in the gut as well, they can have diarrhea and rashes. Malnutrition, failure to thrive, and constant pain and nausea may cause social and developmental delays. It is very important for children with EE to be monitored by feeding and occupational therapists.

Food impaction (food stuck in the esophagus) is the main symptom for teens and adults.<sup>5</sup> A visit to the ER may be required to manually remove the impacted food. EE is not a fatal disease but the side effects of untreated EE can be devastating. The high allergic response of the body, coupled with malnutrition, makes it difficult for a patient to fight infection. A common cold in a healthy person might send an EE patient to the hospital.

Unfortunately, EE is not easy to live with or treat. There is no cure and it is not outgrown with age. There are only two currently accepted treatment plans: dietary changes and steroids. Most doctors favor the dietary route because it has the least side effects.<sup>6</sup> For some this means just cutting out a few foods, such as the top eight allergens. However, most people with EE are allergic to many foods, and some are allergic to all foods. Patients who are not able to eat a balance diet instead rely on an amino-acid based formula. (Amino acids are the building blocks of whole proteins. Broken down, they are not recognized as allergens by the body.) These formulas are foul-tasting and many non-compliant patients end up needing feeding tubes.

Although dietary changes can be extremely effective in sending EE into remission, the social side effects can be heartbreaking. Imagine no birthday cakes, trick-or-treating, or dinner dates. Imagine being afraid to eat food, and getting a tube-feed in the nurse's office while your friends are eating lunch in the cafeteria.

EE is unique in that it is a delayed onset allergic reaction. It is in a different category than common food allergies. Although skin prick and patch testing can be helpful as a guideline for general food allergies, this testing is not generally accepted as a reliable way to determine trigger foods for EE. In a recent study, even those children who did not test positive for food allergies on skin tests showed improvement with their EE by

eliminating common food allergens.<sup>7</sup> Those who went on hypoallergenic formula or eliminated trigger foods had more than a 75% rate of symptom resolution.

The second option is the long-term use of inflammation-reducing steroids. Oral steroids have strong side effects and therefore are prescribed only in life-threatening situations. Fluticasone, a milder inhaled steroid used for asthma, has proven effective for EE when swallowed.<sup>8</sup> It is considered a band-aid patch that does not address the cause for the inflammation. For many children and adults who find it difficult to be compliant with dietary restrictions, it is the most realistic treatment option. There are a small number of EE cases that are triggered by airborne allergens, and for them, long-term Fluticasone is the most logical treatment since dietary treatment would be ineffective.

At the APFED conference I attended, I learned that scientists are testing a new drug called Anti-IL 5 that will inhibit the development of eosinophils in the body.<sup>9</sup> So far the first drug trials are very promising, but taking a pill to manage EE is still years away.

Nitara's pre-existing feeding tube made her treatment choice easy. We put her on the new amino-acid formula for one full month and did not allow her any food at all. Nitara showed marked improvement. Her skin started to show a healthy glow and her hair started to become thicker. She was sleeping through the night and did not vomit nearly as much. Then we introduced some foods that I thought would be safe, one at a time every two weeks. She failed one of them right away: rice. The failure was apparent with symptoms such as vomiting and gagging. Three other foods we trialed she seemed to pass: broccoli, carrots, and potatoes. A biopsy showed good news: she had only 2-3 eosinophils!

Like most parents of children with health issues, I wondered if I did something in pregnancy to cause Nitara's EE. The disease is so "new" that not a lot is known about the causes or triggers. In 2006, Dr. Marc Rothenburg and colleagues discovered a faulty gene on Chromosome 5 that people with EE display.<sup>10</sup> Interestingly, the faulty gene lies right next to the genes thought to cause asthma and allergic rhinitis. EE patients often have siblings or other family members who have the same disease. Parents will often get tested and find they have a form of the disease themselves. This was true in my case--I have a mild form of it and we strongly suspect that my husband's side has the disease as well based on family history. It has been shown that a higher expression of the faulty gene leads to more severe expression of the disease.

Seventy-five percent of people with EE also have asthma, eczema, and allergies or have family members who do.<sup>11</sup> Others, however, appear to have no genetic connection. Certain areas in the United States seem to have higher rates of EE than normal. These "hot spots" may be due to pollution, viruses, or other environmental factors. The normal occurrence of the disease is about 4 in 10,000, although current research is showing it may be more common.<sup>12</sup> In my area, I know of eleven EE patients within a ten-mile radius! Only the most serious cases are biopsied and diagnosed, but milder forms probably exist in higher numbers.



EE may present alongside GERD, hiatal hernias, Crohn's Disease, Celiac Disease, Cystic Fibrosis, and other GI conditions in patients. The relationship between these conditions is unknown.



A healthy Nitara, thanks to a proper diagnosis and treatment

At four years old, Nitara is doing better than ever. She's getting stronger and made it through the winter with no trips to the ER and no antibiotics. She is tall for her age and maintaining a good weight. Nitara is continuing to undergo food trials. She appears to be allergic to slightly more than half of the foods she has tried. Her diet is still extremely limited, but she is currently eating enough foods to just give her a balanced diet. If she shows elevated numbers on her upcoming biopsy, she will lose all of those new foods and go back to formula supplementation. But life goes on. Nitara attends preschool part-time and is outgoing and happy. She loves to sing, take long baths, and wear sparkly shoes. Nitara's relationship with food will always be touchy but her relationship with life looks very promising.

For more information, visit APFED at <http://www.apfed.org/>

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<sup>1</sup> For a general overview of EE, see Glenn T. Furuta, *et al.* Eosinophilic Esophagitis in Children and Adults: A Systematic Review and Consensus Recommendations for Diagnosis and Treatment Sponsored by the American Gastroenterological Association (AGA) Institute and North American Society of Pediatric Gastroenterology, Hepatology, and Nutrition. *Gastroenterology* 2007;133(4):1342-63.

<sup>2</sup> Furuta, 1344-5.

<sup>3</sup> Furuta, 1345-50.

<sup>4</sup> Furuta, 1344.

<sup>5</sup> Furuta, 1344.

<sup>6</sup> Furuta, 1355-6.

<sup>7</sup> Jonathan M. Spergel, *et al.* Treatment of eosinophilic esophagitis with specific food elimination diet directed by a combination of skin prick and patch tests. *Annals of Allergy* 2005;95(4):336-43.

<sup>8</sup> Richard J. Noel, *et al.* Clinical and immunopathologic effects of swallowed fluticasone for eosinophilic esophagitis. *Clinical Gastroenterology and Hepatology* 2004;2(7):568-75; Furuta, 1354-5.

<sup>9</sup> Miguel L. Stein, *et al.* Anti-IL-5 (mepolizumab) therapy for eosinophilic esophagitis. *Journal of Allergy and Clinical Immunology* 2006;118(6):1312-9.

<sup>10</sup> Carine Blanchard, Marc E Rothenberg, *et al.* Eotaxin-3 and a uniquely conserved gene-expression profile in eosinophilic esophagitis. *Journal of Clinical Investigation* 2006;116(2):536-547.

<sup>11</sup> Furuta, 1350-3.

<sup>12</sup> Furuta, 1358.