



Supporting you. Supporting each other.

NATIONAL FOUNDATION FOR
ECTODERMAL DYSPLASIAS

Ensuring Lasting Smiles Act (S.754/HR.1916) Impact on Patients with Congenital Anomalies



This document shares the personal stories of Americans affected by various congenital anomalies whose health would benefit from passage of the Ensuring Lasting Smiles Act. If people such as these go untreated due to health insurance denials or delays, not only will their overall health worsen, but they will be forced to undergo numerous other costly secondary treatments that may or may not improve symptoms and never truly repair the congenital anomaly or restore bodily function. Initial and ongoing medically necessary treatments for congenital anomalies must be approved to improve the health of Americans like these, and that is what the Ensuring Lasting Smiles Act will ultimately accomplish. But they need your support.



Aidan (Wisconsin): After many years of appointments with specialists at Children's Hospital of Wisconsin, a dental resident and a geneticist diagnosed our son Aidan, with x-linked hypohidrotic ectodermal dysplasia (XLHED). This disease affects Aidan's skin, hair, nails, sweat glands, and teeth. His skin and nails are sensitive, and he gets rashes easily. His sweat glands don't function properly, so he has a hard time regulating fevers and overheats easily. He was born without many of his teeth, and has issues due to his lack of teeth, including speech, swallowing, chewing, and eating. He has had to go through invasive procedures, just to determine that his issues are due to his lack of

teeth. When Aidan first started his dental procedures after diagnosis, we were told that since these repairs were medically necessary and needed to be done due to his congenital abnormality, his dental treatments should be covered by insurance. Since his diagnosis, our family has had to fight with our medical insurance for coverage and we soon discovered that very little is covered. Each time we submitted a claim, we have had to appeal several times, and the appeals and denials process for each stage of his treatment is frustrating and unnecessary. Now that Aidan is 16 years old, and most of his dental work needs to begin, we have been told that they have made their final decision to deny his upcoming dental treatments.



Rosie (California): Approximately 1,300 babies born in the US each year have congenital cataracts. My daughter Rosie is one of them. Rosie had surgery at Stanford at six weeks of age with Dr. Scott Lambert. Dr. Lambert has been performing these surgeries for many years and this is his area of expertise (known nationally for his research on congenital cataracts and contact lenses in infants). Following surgery, Dr. Lambert and an infant optometrist fitted Rosie for her first contact lens (glasses are not an option, not powerful enough). My husband and I were shocked to learn these lenses are not covered by insurance (\$200 per lens). The lenses are changed often as the baby's eye grows and vision develops, and the lens can pop out and be lost. Rosie is now 11 months and we have spent over \$4,000 dollars on lenses.

To be clear, without these lenses, these babies are blind and have no chance of ever developing normal vision. As Dr. Lambert likes to say - the analogy is denying a prosthesis to an amputee and telling that patient he or she may never walk again.

If ELSA passes, ensuring that insurance companies pay for these pediatric lenses, then families would not have to make horrific choices such as their child's vision or putting food on the table.



Alli (Iowa): At the age of 6.5 years old, Alli visited her dentist after a playground injury to her top teeth. She had all twenty of her baby teeth and all routine cleaning exam visits had been “normal.” Our dentist took a panoramic dental x-ray as part of her treatment; Upon reviewing the films, he shared the words Ectodermal Dysplasia. We had never heard of Ectodermal Dysplasia, but instantly knew it would become a big part of our lives. We spent the coming months and years commuting 240 miles round trip to University of Iowa Hospitals and Clinics. Appointments with genetics, dermatology and several dental specialists resulted in a clinical diagnosis of Hypohidrotic Ectodermal Dysplasia (HED). It was identified

that Alli had 12 permanent tooth buds, which meant she would be missing 20 adult teeth. Learning more about HED, we finally had clarity about her gastrointestinal issues, inability to eat certain foods, frequent overheating, pigment issues with her skin, brittle nails and dry hair. Currently Alli is two years in on her second set of orthodontic braces, as soon as her braces are ready to come off, she will be ready for her first dental appliance. This will not be covered by our medical insurance even though the cause of her missing teeth is a birth defect and causes several other health related issues.



Lauren (Tennessee): Hi! My name is Lauren. I was born with a bilateral cleft lip and palate which means I had splits on both sides of my upper lip leading up to the base of my nose and I was lacking a roof in my mouth. By the time I was 7 years old, I had seven surgeries to help correct my cleft lip and palate. Many of these surgeries were considered cosmetic because they were performed by a cosmetic surgeon and insurance would only cover some – but in some cases none – of the costs. I wore braces just short of ten years and went to speech classes for three years to overcome lisps and compensating pronunciations. At age 17, I had another major surgery which consisted of surgically repositioning my upper and lower jaws and chin to correct a severe underbite and to add support in the upper parts of my face

(i.e. cheek area) that I was congenitally lacking. My health insurer initially deemed the surgery unnecessary and notified me that I would have to pay for it myself. My oral and maxillofacial surgeon pressed back against the insurer explaining that if I did not have this surgery, it would impair my speech that I had worked so hard to correct, my face would slowly concave in and grow more in a “c” shape and my underbite would get progressively worse making it harder for me to eat everyday food. The insurer ultimately agreed to cover the cost of surgery; however, they subsequently denied coverage for a dental-related procedure associated with my condition. Due to the hole that originally existed from the top of my lip to my nose, I had a missing front tooth and needed a permanent prosthetic tooth, or a “dental implant.” I applied to my health insurer to help cover the procedure, but because the placement of my absent tooth was in the front and not a back molar they considered replacement of the front tooth “cosmetic.” Tell me having a front tooth is not essential to securing a job, eating, or having self-esteem? I had to borrow \$13,000 to cover the cost of that procedure. Overall my parents and I have spent \$20,000 to cover the cost of my treatment since birth. While I am beyond grateful that my family

and I were ultimately able to pay for the various stages of my treatment, it did not come without ripple effects. The expenses we paid to help me speak, breathe and eat – which allowed me to go on to become a pediatric nurse – have placed unnecessary financial strain on me and my parents not only in the present, but for the years to come.



Caleb (Rhode Island): My name is Caleb. I'm 9 years old, and my wish is to have a full set of teeth. I have 4 upper teeth all misshaped, 2 of which are centrally located but not fully erupted. My dental team consists of a maxillofacial surgeon, prosthodontist and orthodontist. My x-linked hypohidrotic ectodermal dysplasia has left my lower jaw short and thin throughout. I have no ridge for even a denture to rest on. There is almost no bone in my posterior upper jaw and both the height and width are significantly deficient. My short-term goals require me to have a sinus augmentation to add bone taken from my hip and implanted between my jaw and sinuses. This will

allow me to have upper implants. My long-term goals would include another bone graft to reconstruct my bottom jaw allowing me to receive implants. Timing...due to my age, my team is concerned about psychological and social influences while my Mom and Dad are concerned of the high cost of medical care required due to my health insurance not recognizing my oral care as a congenital anomaly. Your concern to close the loophole in my insurance will make my wish come true.



Karl (Minnesota): My name is Karl. I was born in rural Minnesota and wasn't diagnosed with ectodermal dysplasia until the age of 5 when my dental challenges became obvious. I have only a few baby teeth and even fewer adult teeth. I received my first set of dentures when I was 5 and they quickly became an amazing tool for "shock value" during my grade school years! As I grew, I needed larger dentures and my parents struggled to get them covered by our medical insurance, despite numerous appeals as the dental needs were a result of

my medical condition. During my 20s, my teeth deteriorated and were not viable to support another denture. I had to make a hard decision and I was advised to pursue dental implants at the University of Minnesota. This was a great decision and the right treatment plan for me. The implants and associated prostheses were both an improvement in function as well as appearance. However, it came with a significant price which I had to pay out of pocket as my medical insurance wouldn't cover it. 10 implants, a fixed removal denture on the bottom and a removal denture on the top cost me well over \$30,000 in 2000. It was a hard decision, but it was the right decision.

In my mid-40s, a couple of the implants in my mandible failed. Bone grafting was needed to build up the site for restoration again. They were able to shave off the top margin of the back of my mandible (ramus) to harvest enough material to fill in the defect. It took several months before the bone was ready for a new implant to be placed. I have been advised to replace the implants in the maxilla soon. During this phase, I'll need bone graft material to be placed in my sinus cavities to help anchor the replacement implants since my bone density is abnormal, like many people with ectodermal dysplasia. As you can imagine, I'm not looking forward to this step.

My medical insurance does not cover any of the implants or the new dentures that I'll need for my oral restoration even though I work hard and pay my premiums. Please support this legislation so medical insurance companies clearly understand that teeth are not cosmetic and that restorations of congenital

anomalies, including dental changes, should be included in their medical plans. Let's get this right so my daughter doesn't have to be fighting for every single tooth. Let's get this right so your daughter, your son, your granddaughter, or your grandson doesn't have to repeatedly read denial letters.



Cody (New Mexico): Hello all. I'm Cody. I'm 33, a husband and father of two. I have X-linked hypohidrotic ectodermal dysplasia and my daughter is an affected carrier. I had a total of six teeth, and all were conical. I was never able to eat the same things others enjoyed: jerky, nuts, steak, apples. I just couldn't chew them. I was fitted for dentures several times in childhood and they didn't help. Not to mention the unnatural look and embarrassment of caring for them. At one point, I was fitted with dentures that were glued in to prevent the constant care of a 6-year-old using Poligrip just so he could eat. After various tries, I threw in the towel and just didn't eat what others could.

As an adult, I began the process of finding a maxillofacial surgeon and dentist to install dental implants. There were none local, so I had to travel 3 hours for consultations and appointments. I had orthognatic surgery taking bone grafted from my hip to align my jaw correctly. The bone that would typically hold tooth roots or implants had deteriorated from the lack of teeth and had to be built back up with my bone grafted from my hip. A total of 10 screws were used in my jaw to align and attach the new bone. This 12-hour surgery cost \$15,000 out of pocket for the surgery alone. The follow-up process took 6 months to heal.

The next step was removing my teeth. This was the most nerve-racking part. I only had 6 teeth to begin with and now I was going to have zero. This process was \$1,800 out of pocket. I then had dentures fitted and installed with my dentist to get by, costing \$1,500. After three months of healing, the doctor installed the actual implant posts at \$7,000 out of pocket. After three months, the doctor uncovered the implants. My dentist took molds and measurements to build the actual teeth at a cost of \$7,000. After 18 months and \$30,000, I had a mouth full of semi-permanent teeth.

Since my teeth were completed, I have spent around \$500 a year in appointments and repairs. The teeth have broken, the abutment gaskets wear down and most recently, my complete upper broke into pieces. The brand of abutments the dentist chose to use on the bottom were subpar, so they don't stay in place. Fixing these two things alone will total over \$7,000 and I continually get denied by insurance. I received a bill just for taking the broken upper out last week and it's \$400 and the insurance is not going to cover any of it. My daughter is 4 years old and we will begin going through the same process for her. Thankfully she has twice as many teeth as I did but the majorities are also conical so she, too, will have the same eating difficulties I have had.