

Meet Peter Dankelson

September 2010

Goldenhar Spectrum is a birth defect (about 1 in 5,000 live births) characterized by abnormal prenatal development of the head and face. Common anomalies include missing ears and malformations of the jaw, eye, vertebrae, mouth and palate. Asymmetry is also a common characteristic with one side of the body more affected than the other. Other names used for Goldenhar Spectrum include Oculo Auriculo Vertebral Spectrum (OAV) and Hemifacial Microsomia. Scientists do not know what causes Goldenhar, however, there are strong indicators that it is more environmental vs. genetic.

Peter's Goldenhar anomalies at birth included microtia/atresia of left ear (missing left ear and no ear canal), dermoid cysts in both eyes, cleft palate (roof of mouth), micrognathia (underdeveloped lower jaw), absence of TMJ joint on left side, right single pelvic kidney, cervical and thoracic spinal fusions, hemi-vertebrae, and two extra ribs on his left side. Peter was trached at birth due to obstruction of his airway. His jaw was so small that his tongue fell back in his throat and cut off his ability to breathe through his mouth or nose. He was decannulated (trach removed) on October 21, 2004 (about two weeks before his baby brother arrived) but **still has an open trach stoma** (hole in his neck) and an extremely small airway.

Due to his jaw abnormalities, Peter was not able to eat orally for the first 2+ years of his life. He **still uses his feeding tube** for medications and supplemental nutrition. In addition to his Goldenhar anomalies, Peter was born prematurely at 30 weeks gestation on October 7, 2000. His birth weight was 2 pounds 13 ½ ounces. Peter **has hypertension**, which is controlled with medication; he **wears an auricular (ear) prosthesis on his left side** and glasses to correct astigmatism and nearsightedness.

Peter is currently nine years old and has had several reconstructive surgeries including repair of cleft palate, bone graft (using rib bone) to left mandible, mandible distraction of lower jaw, removal of dermoid in left eye, removal of bronchial cysts on left side of face, placement of a tube in his right ear, and multiple bronchoscopies (scopes of his airway). He will probably need 1-2 more jaw distractions to improve his airway as he grows. Several years from now, he will have a major surgery to reconstruct his trachea and close the hole in his neck.

Peter has annual x-rays taken of his neck to check for instability and curvature of his spine. Because of this, his orthopedist has **restricted him from doing anything that puts direct pressure on his head or neck. He is also restricted from playing any contact sports due to his single-pelvic kidney. Because of his open airway, he cannot go underwater.** He is followed by about eight pediatric specialists.

Peter is evaluated every 6-12 months by audiology and has normal hearing on his right side. He sometimes wears a digital sound processor on a headband to help him hear. He is cognitively normal, and he currently attends 4th grade at St. Patrick Catholic School in White Lake, Michigan. Peter loves to read, play video games, watch movies, and play with his younger brother, Jacob.



Peter at one and seven years old