

## Matters of the Heart

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**One family's struggle to honor their children will comfort those coping with similar misfortunes and help to find a cure for a rare syndrome.**



Laurian and John Scott, former Phoenix residents, were not new to facing extreme obstacles or learning how to balance fear and emotion. Their newest arrival, Noah, would undergo open-heart surgery at just two days old. What the Scotts were not prepared for was the silent disease that would first overpower their daughter, Elenna "Thisbe," and then take aim for Noah.

It began on Thanksgiving of 2005, when Laurian noticed a slight wheeze from then 17-month-old Thisbe. Attributing it to allergies or a cold, Laurian was not alarmed. Over the next week, Thisbe's slight wheeze progressively grew worse. Doctors at Phoenix Children's Hospital thought Thisbe's wheeze stemmed from a swallowed object and labeled her wheeze a "stridor" (a sharp sound heard only on an inhale). X-rays were inconclusive, so doctors went forth with surgery. Tests soon confirmed that Thisbe's vocal cords were paralyzed. On New Year's Eve, Thisbe was rushed to the hospital, where she turned blue. That night, the Scotts made a wrenching decision and agreed with doctors that Thisbe needed a tracheotomy. That decision would leave Thisbe with no vocal ability and Laurian and John without hearing their little girl's voice again. Five exhausting months later, Thisbe was diagnosed with Brown-Vialetto-Van Laere Syndrome (BVVL).

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On Thanksgiving of 2006, exactly one year later, Thisbe stopped breathing. "[She] died in our arms," Laurian recalls. Although the Scotts were able to resuscitate her, they knew Thisbe's illness had taken a turn for the worse. "The last eight months of Thisbe's disease were worse than any nightmare," Laurian says. "I would wake up and realize I would so much rather be asleep in my nightmare than awake in this reality."

Elenna "Thisbe" Scott died on April 30, 2007 at 21 months old.

One month after Thisbe's death, Laurian and John were not done living the nightmare. They soon discovered Noah's stridor. In July 2007, Noah, just 10 months old, showed an initial sign of BVVL in the form of a weakened eyelid. Loss of hearing and vocal paralysis soon followed. As the disease ravaged Noah's body, Laurian and John made the gut-wrenching decision not to go ahead with a tracheotomy, knowing it would only prolong the discomfort. The Scotts held out hope that Noah's course would be different than Thisbe's. A gut feeling told them different.

Noah Scott died April 9, 2008 at 19 months old. Thisbe and Noah leave behind a sister, Aslan.

Brown-Vialetto-Van Laere Syndrome, a debilitating disease that can affect anyone from infants to adults, is a variant of Motor Neuron Disease. In addition to the loss of mobility caused by MND, Brown-Vialetto-Van Laere steals one's hearing. Dr. Brooke B. Allen, a physician-neurologist on the advisory board of BVVL International and Thisbe's godmother, says this disease attacks the motor neurons that are responsible for the basic function of life. "Every motor neuron disease will rob a person of their body and keep their mind intact," Laurian says. To date, there have been only 58 reported cases of BVVL worldwide since the 1800's. Thisbe and Noah will be the 59th and 60th documented cases.

To commemorate their children's brief lives, the Scotts founded The Olive Branch Fund, a nonprofit that is a component of The Community Foundation of Middle Tennessee. The two foundations partner with Genetic Alliance in hopes of supporting the search for a BVVL cure. "The only problem with rare diseases that we have learned, is they are rarely documented," Laurian says.

Recently, two U.K. clinicians narrowed the search to 80 possible genes. Further funding and research will expedite the research process. Thisbe and Noah's case documents will aid in future research.

"If we can find a gene, that is huge," John says.

This month, the Scotts will host "A Mighty Voice" fund-raiser at the Phoenix Zoo benefiting The Olive Branch Fund: a Thisbe and Noah Scott Legacy. The fund-raiser will also collaborate with the release of Laurian's picture book, a tribute to Thisbe and other BVVL-affected families with a message "to never give up hope."