

Listening to a Hunch

Children's doctor follows his intuition and opens a new door that may reveal a secret to SIDS.

NEARLY 40 YEARS HAVE PASSED SINCE CHILDREN'S HOSPITAL HOSTED AN INTERNATIONAL CONFERENCE THAT SHINED A BRIGHT LIGHT ON A DARK TOPIC.

In 1969, Sudden Infant Death Syndrome (SIDS) — lacked a name, a definition or even acceptance as a distinct diagnosis before the conference. The mysterious death of a seemingly healthy infant in his or her sleep was simply a rare tragedy.

While SIDS remains tragic and mysterious, SIDS deaths are less common today. The conference and the leadership of Dr. Bruce Beckwith — who began studying crib deaths in 1964 — helped illuminate SIDS and bring about greater understanding of how to reduce its occurrence.

It was Beckwith, former head of pediatric pathology at Children's, who coined the name SIDS. "We didn't discover the disease," recalls Beckwith, "but we were the ones who gave it a name and made it an internationally recognized problem and inspired other researchers to begin looking at it."

Four decades later, much has been learned about SIDS, yet the quest for its cause continues. At Children's, where so much of the initial investigation of SIDS occurred, Dr. Daniel Rubens is following in Beckwith's footsteps.

In a study published earlier this year, Rubens found a strong connection



Though he had no previous background in SIDS research, Dr. Daniel Rubens followed a hunch that could lead to a new understanding of SIDS.

When seeking information about newborns' inner ear function, Dr. Daniel Rubens turned to audiologist Susan Norton, PhD, who has done extensive research on newborn hearing.



between SIDS and an abnormality in the ear. It's a potential breakthrough, but also an example of how medical research is a journey where opening one door leads to another and where unrelated paths often cross in unexpected ways.

Intuition inspires search

Rubens is an anesthesiologist, not a SIDS expert, yet his experience dealing with infants in crisis inspired him to seek an answer to the unsolved riddle of SIDS.

Rubens listened to his intuition when he decided to seek clues to SIDS in the inner ear. "I had the idea that we've missed something in medicine about the way the body controls breathing and that the missing piece might be in the specialized nerve tissue found in the inner ear," he says.

When Rubens sought hearing-related data to pursue his theory, he turned to Susan Norton, who leads Clinical and Research Audiology at Children's. Between 1993 and 2000, Norton led a national study to compare the effectiveness of various newborn hearing tests.

While leading the study, Norton worked closely with Dr. Betty Vohr of Womens and Infants Hospital, in Rhode Island, one of the first states to test the hearing of all newborns. Norton's relationship with Vohr paved the way for Rubens to gain access to 13 years of Rhode Island hearing test results.

The Rhode Island data was a gold mine for Rubens. The state's testing

program was able to provide a large enough sample of SIDS cases — 31 — to be statistically significant. And the sensitivity of the tests enabled Rubens to detect a hearing deficiency in the right ears of all 31 babies, each of whom showed a consistently lower score across three sound frequencies when compared to babies who did not die from SIDS.

Much follow-up remains to be done. However, the finding raises the possibility that hearing tests might be used to screen newborns for risk of SIDS — which typically occurs between the ages of 3 weeks and 6 months — and that the screening results might allow doctors to intervene before tragedy strikes.

"It's a preliminary finding, but it's too strong to ignore," says Rubens. "We never thought to look in the inner ear. It points us in a whole new direction."

Deadly CO₂ buildup

Rubens believes that babies who die from SIDS suffer some sort of trauma at birth that damages the fine hairs of the inner ear. That would explain the hearing loss, but Rubens suspects some of those hairs also signal the brain about carbon dioxide (CO₂) levels in the blood.

Normally, the brain would respond to mounting CO₂ levels by increasing the rate and depth of breathing. Rubens suspects SIDS is caused by damage to the fine hairs, which prevents the hairs from warning the brain about CO₂ levels and adjusting breathing accordingly.

The danger multiplies during sleep, when breathing already is slower and shallower than normal. "Basically, the babies suffocate because they're not getting enough oxygen," hypothesizes Rubens.

While the link between fine hairs in the inner ear and respiratory control remains unproven, data from the Rhode Island hearing tests gave Rubens a way to connect the first dot and associate SIDS with an abnormality of the inner ear.

"Nothing ever happens in isolation," says Norton. "Daniel could not have begun to ask the question without the data from Rhode Island."

Picking up the baton

Rubens also drew upon the legacy of Beckwith. "It's been my privilege to follow in Dr. Beckwith's footsteps," he says. "Each new breakthrough brings us closer to making SIDS a condition of the past."

The most important SIDS advances so far involve preventive measures — especially the knowledge that babies should not sleep on their tummies.

Over the years, Beckwith has seen many promising theories about the cause of SIDS fail to pan out. "It's a tough nut to crack," he says. And, whether Rubens is on the road to pinpointing the cause of SIDS remains to be seen.

"His research is definitely worth following up on," says Beckwith. "It may be a misleading finding, but it could be an important clue that could help us identify babies at risk."

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