

By Ione Echeverria

**Like most parents,**

Tammy Stewart enjoyed taking candid photographs of her 9-week-old daughter, Alexis Diaz. But a particular set of pictures made Stewart uneasy.

“In some of the pictures, Alexis’ right eye had a golden, iridescent reflection, like a cat’s eye,” said Stewart, a stay-at-home mom from Duncanville. “I showed the pictures to family and friends, but they all said I was overreacting.”

Trusting her instincts, Stewart took her daughter in for another examination. Alexis’ pediatrician used an ophthalmoscope — a lighted instrument used to examine the interior of the eye — to elicit a red reflex from her retina. In Alexis’ left eye, the reflex was normal. But her right retina appeared white, a sign that there was an abnormality.

“Red-eye,” which can be captured in a photo, is caused by a reflection off the blood in the retina. Leukocoria, or white pupil, is caused by something blocking the reflex. Absence of blood in the retina would have to be extreme to reduce the coloration.

Alexis was referred to Dr. Albert Edwards, assistant professor of ophthalmology at The University of Texas Southwestern Medical Center at Dallas. Tests revealed that the infant girl had retinoblastoma, a rare cancer of the retina.

Each year 350 American children are diagnosed with the disease, which can occur in unilateral (tumors in one eye) or bilateral (both eyes) form. UT Southwestern ophthalmologists treat a handful of these cases at the James W. Aston Ambulatory Care Center and at Children’s Medical Center of Dallas.

“Retinoblastoma is a childhood disease that is typically diagnosed before age 5,” said Dr. Nick Hogan, assistant professor of ophthalmology, neurology, neurological surgery and pathology. “Although the disease is rare, it is the third most common cancer and the No. 1 ocular cancer affecting children.”

Retinoblastoma, however, can now be cured in 90 percent of cases because it can be detected early and treated aggressively.

Children with the hereditary form of retinoblastoma typically develop the disease at a younger age, have bilateral eye tumors and an increased chance of developing other cancers in later years.

**A picture is worth a thousand words**

When retinoblastoma was detected in Alexis’ right eye, Edwards ordered a battery of eye scans and tests, including a systemic work-up to determine if cancer was present in other parts of her body and brain. Based on the results, her eye was removed in a procedure called enucleation.

“Conservative treatments, such as radiotherapy and chemotherapy, have decreased the frequency and necessity of enucleation, but it still is necessary in some cases,” Hogan said.

Doctors and patients must weigh the pros and cons of different treatments, Edwards said.

“If the eye has large-sized tumors that fill up 50 percent of the back of the eye or is fragmented, the eye is typically removed because its survival is not likely after external beam radiation treatment,” he said.

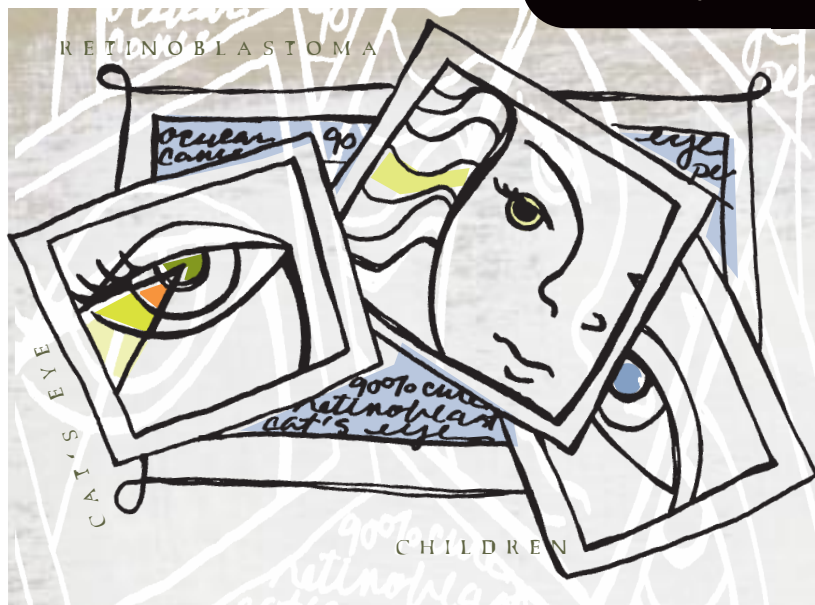
At the time of Alexis’ surgery, an adult-size anchoring implant was placed in the eye orbit and muscles were reattached. Because the implant is made of a derivative of coral, it is porous enough to allow blood vessels to grow into it. The little girl was then fitted with a cosmetic prosthesis that matched the color of her other eye. The prosthesis will continue to be refitted as her healthy eye grows.

Looking at Alexis, who is now an active and happy 3-year-old, Stewart is grateful that her daughter’s life and eyesight were spared.

“Alexis started pre-kindergarten this fall,” Stewart said. “Despite all the poking and prodding she’s had to endure in her young life, she has adjusted well to living with one eye.”

And she’s ready for her first school photograph.✻

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—Tammy Stewart



By Amy Shields

## The heart pumps

about 1 million barrels of blood during an average lifetime. But for the 5 million Americans suffering from congestive heart failure, the organ's ability to perform this monumental task is greatly diminished.

A new Food and Drug Administration-approved pacemaker, however, is offering new hope for these patients.

The University of Texas Southwestern Medical Center at Dallas is one of the first sites in Texas to offer the biventricular pacemaker. The device resynchronizes an uncoordinated beating heart with electrical abnormalities, allowing the heart to more efficiently pump the 31 million gallons of blood circulated during an average lifetime.

In about one-third of people diagnosed with congestive heart failure, the left ventricle — the heart's main pumping chamber — is no longer synchronized. As a result, the heart cannot push blood out to the rest of the body as efficiently. This leads to fatigue, complications of the respiratory system and, sometimes, death.

The new pacemaker significantly reduces the complications associated with congestive heart failure, improves a patient's mobility and cuts hospitalizations by almost half, according to a study published in the June 13 issue of *The New England Journal of Medicine*.

## Setting the pace in a heartbeat

"The biventricular pacemaker is designed for persons with severe congestive heart failure who are not responding to optimized medications and have electrical conduction delays in their hearts," said Dr. Robert Kowal, assistant professor of internal medicine who performs the implantation procedure. "It provides a non-

pharmacologic method of improving heart function, making patients feel better and allowing them a greater exercise capacity. In the long term it may decrease mortality from congestive heart failure."

A standard pacemaker addresses rhythm disturbances. The biventricular pacemaker is designed to treat impaired blood flow by resynchronizing the ventricles of the heart. Implanted in a patient's chest during a minor surgery, the device simultaneously paces both the right and left ventricles.

"Preliminary studies have shown that biventricular pacing may decrease hospital admissions by up to 70 percent in these patients. I think that is incredibly promising," Kowal said.

Although the pacemaker improves quality of life, the device is not designed to replace drug therapy to treat the condition, said Dr. Clyde Yancy, medical director of the UT Southwestern/St. Paul Heart Transplant Program.

There are several causes of congestive heart failure, including coronary artery disease, heart valve disease and hypertension. General treatment includes salt restriction, diuretics to get rid of excess fluid, digoxin to strengthen the heart and other medications.

Kowal and other cardiologists in the electrophysiology lab at Parkland Memorial Hospital began implanting biventricular pacemakers in October 2001. Similar devices are being implanted at the Dallas Veterans Affairs Medical Center where Dr. Mohamed Hamdan, associate professor of internal medicine and holder of the Dallas Heart Ball Chair in Cardiac Arrhythmia Research, is leading research in this area.

"There are some reasons to suggest that this type of pacing will not only improve the heart's pumping ability and allow the heart to pump more efficiently, but may decrease the chance of people having life-threatening heart rhythms and limit the number of people who need transplants," Kowal said. "I think it's going to be a real breakthrough in treatment options for congestive heart failure."\*

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—Dr. Robert Kowal



P A C I N G P U M P I N G

By Ann Harrell

**Sharon Hahs had no idea** a tumor that appeared in her neck was a late sign of ovarian cancer.

“Three-quarters of women with ovarian cancer present with their disease at an advanced stage because they have no symptoms, and there’s currently no accepted test to detect it early enough to prevent its development,” said Dr. John Schorge, assistant professor of obstetrics and gynecology at The University of Texas Southwestern Medical Center at Dallas.

Currently, the disease is fatal for about 56 percent of the patients diagnosed with it, making it the most deadly of the gynecologic cancers. However, good news may be on the way.

Schorge and his team of specialists, along with medical scientists from Harvard Medical School and three other academic medical centers, last spring found a marker for ovarian cancer that utilizes osteopontin, one of the body’s many proteins. Osteopontin, which may be revealed by a simple blood test, is found in bodily fluids such as blood plasma, urine, milk and bile. It is elevated in ovarian cancer patients, even those in the early stages of the disease, leading researchers to believe they are on the way to developing the first successful early-stage ovarian cancer test.

Most of the patient research was conducted at UT Southwestern under Schorge’s supervision, and his findings were published in the April 3 issue of *The Journal of the American Medical Association*.

There is currently only one blood test associated with ovarian cancer, Schorge explained, but it’s not effective as an early diagnostic tool. The test, called CA125, is used to follow women who already have been diagnosed with the disease to determine whether they are responding to treatment.

“We identified a specific protein, osteopontin, that is overproduced 184-fold in ovarian cancer cells. Then we wondered if that was measurable in the blood. So we tested it,” said Schorge.

Preliminary results of the first stage of testing showed that osteopontin levels in blood plasma were significantly higher in 51 patients with epithelial ovarian cancer than in 199 other women, including 107 healthy controls, 45 patients with benign ovarian disease and 47 with other gynecological cancers. The researchers recommended further testing because of the early success with the biomarker.

## Overtaking early ovarian cancer

“Every year 25,000 patients with ovarian cancer are diagnosed, and approximately 14,000 of these women

die,” said Schorge, who is a member of the Harold C. Simmons Comprehensive Cancer Center team at UT Southwestern. “That’s why finding a biomarker that can be used at the beginning is so vital.”

Schorge explained that most ovarian cancer is detected at Stages III or IV, the most advanced levels of the disease. Patients with Stage III or IV ovarian cancer have a five-year survival rate of less than 30 percent while patients detected during Stage I have a 95 percent survival rate. Medical scientists have investigated a number of markers for ovarian cancer, he said, but none so far have been successful in detecting early-stage disease.

“Our team is hoping that the osteopontin, which is present in the actual tissue of the cancer, will lead to the development of an early blood test for ovarian cancer that could affect the lives of all women,” Schorge said.

The next step will be a clinical trial using a variety of early detection biomarkers, including osteopontin.

Within five years, Schorge said, there could be an economical blood screening test for ovarian cancer that might be used annually in conjunction with a Pap test for cervical cancer.

That’s why Sharon Hahs volunteered to be part of the ongoing UT Southwestern study.

“If you know you have ovarian cancer already, but the doctors are working to catch it early, other women may have a better survival rate,” Hahs said. “Anything I can do to help that research I’m willing to do. I want to be part of the solution.”



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**—Dr. John Schorge**