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## **New medications, 'rodding' offer hope for child afflicted with brittle bones**

By Dolores Harrington

Second of two parts

When Isaiah Vining was diagnosed, shortly after his birth, with osteogenesis imperfecta (OI), his parents, Lon and Amanda, had no idea how serious his condition might be or what his prognosis was.

OI is also called "brittle bone disease," and those who have it are subject to frequent fractures. It often includes loose joints and muscle weakness.

OI has been classified by several types with varying degrees of severity. However, the symptoms in individuals diagnosed with the same type of OI are often different. It appears that Isaiah's symptoms fall between types III and IV -- one of which is referred to as "progressive deforming" and the other called "moderate severe."

Type II OI is considered "lethal," and frequently leads to death at or shortly after birth. Type I is considered mild. Lon Vining said, "Our specialist says they really don't have a good definition of what OI is and what it isn't."

There is help for most children with OI. "They have a drug that helps build bone mass," Lon said. Isaiah is given the drug, pamidronate disodium, which is one of the drugs used for persons with osteoporosis (thinning bones.)

The drug is not approved for use in children, but doctors can prescribe it for them. Isaiah is given the drug intravenously every eight weeks, and he has a port beneath his skin to avoid the pain and problems connected with starting an IV for each infusion. "You can tell when he's had his medicine," Amanda said. "It helps his pain."

Lon said X-rays show where he's had the drug, "It's like rings on a tree. It appears, by his bone scans, that he's not getting any worse. He may be getting better."

Another recent development is the invention of Fassier-Duval rods. The rods are inserted into the bones -- the procedure is called "rodding" -- to keep them straight and help prevent fractures; OI bones tend to bend or bow. The rods, named for the Canadian inventors, are made to expand with growth -- perhaps for as long as three years.

In May 2004, Isaiah had the rods put in both femurs -- thighbones. He was the youngest child to have the rods in the femurs. Last month, he became the first child in the U.S. to have one of the rods in a tibia -- the inner bone between the knee and ankle.

The surgeries to implant the rods are done in Omaha, Neb., the first place in this country where the procedure was done. The surgery on Isaiah's tibia was performed on Jan. 21, and he and his parents came home on Jan. 26.

"I took him to Dr. McLeod to get his cast off," Lon said. Kevin McLeod, a local orthopedic doctor, thought it was too soon. It had only been a little over two weeks since the surgery, but the cast was removed, according to the Nebraska surgeon's instructions.

Exercise is essential for bone development, and children with OI need to be able to exercise as much as possible. Having limbs in casts for long periods of time is detrimental to the development of the bones. The Vinings have learned to make some of the decisions for Isaiah's care. "We've had to become our own specialists," Lon said.

When Isaiah breaks, he is given medicines for the pain and Valium to stop the spasms that occur. At first doctors were reluctant to give the child the tranquilizing drug. For one thing, they had no idea what dosage was safe for an infant. When Isaiah's parents insisted that he have it, the doctors sought and found the answer to how much to give him.

The periods following Isaiah's surgeries or fractures are the most difficult for his family. He must have pain medication and be kept quiet. He is, of course, less active for a while. "Due to the trauma of the fracture and the sedative effect of the pain medications, it's like we lose our child for two weeks," Lon said.

As difficult as it is, Isaiah's family has learned not to restrict him unnecessarily. "The carefulness is the emotional part," Amanda said. "For a while, he was with me all the time. I just couldn't let him go to anyone else, and truthfully a lot of people were scared to take him from me for fear of breaking him."

Amanda has begun to let go a little, and Isaiah spends some time at Pediatric Specialty Care, where he receives a variety of therapies. His fractures and the ensuing recoveries have kept him behind developmentally. He learned to walk last December at 18 months, but on Jan. 9 he fractured his right tibia for the fourth time -- the third time in six months.

He hasn't been able to walk again since that fracture and the rodding surgery in January, but he does get around by crawling. Amanda said it's so exciting for a mother to see her child walk and reach other milestones for the first time, and, "I get the joy of it over and over again."

The Vinings have become active with the Osteogenesis Imperfecta Foundation (OIF), based in Gaithersburg, Md. Lon said the organization is only 20 years old, and "we've only had paid staff for eight years."

He said, "The OIF provides support, education and information to those who have OI and to the general public. It does research for finding treatments and a cure for OI, but compared to most other disorders, OIF is underfunded." The foundation will have only \$400,000 for research this year, and that almost exclusively from gifts and fund-raising.

It is estimated that there are between 20,000 and 50,000 persons with OI, a small number compared to some other diseases and conditions. "There are so few people who have OI," Lon said, "that not enough research has been done."

He said there is a lack of research on the part of the government, although the National Institute of Health does some. "The pharmaceutical companies haven't done much research, because there wouldn't be enough people to sell drugs to -- to make a big profit."

Last summer, the whole Vining family attended a national conference of the OIF. They became acquainted with a group of four families, each of which had a member with OI. "Three out of the four with OI had fractures while we were there. Isaiah had the last one," Lon said. During the final speech, "Isaiah fell on his leg and broke a femur."

To help fund the OIF, the Vinings have established the Isaiah Vining Foundation for Children with Brittle Bones. Donations can be made at Regions Bank, and all donations will go to OIF causes. They are also planning some local fund-raisers and are helping the foundation with various projects to raise money.

They have become acquainted with other families of OI victims via an Internet Web site, OI Parents Yahoo Group, and they've heard some horror stories from other parents. OI can be difficult to diagnose, and it takes about two months to get the results of genetic testing.

Because of that fact, "many parents have been accused of child abuse," Lon said. In one case, the mother of 10 healthy children was accused of abusing her 11th child when the child suffered broken bones. Lon said the only way an otherwise healthy 3-month-old can suffer broken bones is through negligence or abuse. "He can't harm himself under normal conditions."

When doctors and law enforcement officials find a young child has suffered several broken bones, their opinion is that the child has been abused. The child is frequently removed from the parents' custody.

Lon said that sometimes even after OI has been diagnosed, another reason is found not to return a child to his parents. He and Amanda are convinced that many of the accused parents they've been in contact with are innocent and that their children have OI.

There was no doubt that Isaiah had something wrong at birth. "He was clearly broken in utero and during birth," Lon said.

Isaiah will be 22 months next Saturday, and he's had 27 broken bones, including a skull fracture, broken arms and numerous breaks in his legs. His parents don't ask "why Isaiah?" or "why us?"

"Everybody is given a life to live. We figure God has a specific life for Isaiah and maybe for all of us. If God didn't spare His own son ....," Lon said.

Amanda said, "God gave us a child we can take care of. It's a privilege. He's a blessing. He's a happy child. He helps us."

On a recent warm afternoon, Isaiah sat at the end of the walk in front of his house. He and the family dog shared a couple of cookies, and when he was ready Isaiah crawled up the walk and in the front door.

He sat down, turned around and pushed the door closed. And grinned at a visitor.