

PROTECTED TOMORROWS

POLAND'S SYNDROME

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My Story--Introduction

I was keeping an eagle eye on my pre-schooler that summer day at the gorge. Sharp rocks, swift water, stinging insects—so many dangers for an overprotective mother to worry about. But cause for concern was about to come from a totally unexpected place.

Amir had come out of the water and was shivering as his dad dried him off with a big towel. Suddenly my husband called to me with concern in his voice.

“Look at Amir’s chest! What’s wrong with it?” he was asking me.

I looked at my shivering son. The right side looked flatter, more caved in than the left side. The right nipple was higher up on his chest than the left. It wasn’t red or swollen, however.

“We have to take him to the emergency room,” my husband was saying. He was already packing up the blankets and picnic gear. I wasn’t sure it was an emergency situation. But could Amir’s chest have been like that for a long time without my having ever noticed it?

Our next-door neighbor was a surgeon and good friend, so I convinced my husband to call him when we got home. He examined Amir and said he had Poland’s Syndrome, a congenital defect. He made Amir put his hands in prayer position and showed us that the fingers of his right hand were smaller than those of his left.

“Amir has had this since birth,” the doctor told us. “It’s very rare; your pediatrician quite possibly is not even familiar with it. He doesn’t have a pectoral muscle on the right side; that’s why his chest looks so flat.

“Amir’s case is not too severe. Don’t feel bad about not having noticed it. There’s nothing you could have done to prevent it, and it’s not a disease. It’s not going to get any better or worse.”

What is Poland’s Syndrome?

Poland’s syndrome is a rare disorder, characterized by deformities of the chest and hand on one side of the body. It is named after Sir Alfred Poland, the British surgeon who first described its occurrence in 1841. Poland’s syndrome is sometimes known as Poland’s Anomaly. It is also sometimes written “Poland” rather than “Poland’s.”

Poland's syndrome is thought to occur in between 1:10,000 and 1:100,000 births. Gathering statistical data is difficult because it is so uncommon, and because mild cases may go unreported.

What causes Poland's Syndrome?

The condition is thought to be the result of a tiny stroke occurring around day 48 of the life of a fetus. At 6½ weeks after conception, an unborn child is developing limbs. The fingers are beginning to separate, and nipples are forming. Momentary disruption of the blood flow to one part of the body causes a variety of defects.

Is it hereditary?

Exactly why this mini-stroke happens is not known. It has been suggested that there might be some hereditary predisposition towards its occurrence. Cases have been reported of a mother and daughter, or father and son, with similar manifestations of the syndrome. These cases are rare, however. There are also cases studies of twins, one with Poland's syndrome and one without. Some experts have also theorized that Poland's Syndrome may be the result of exposure by the mother to toxins before birth, but this is only a theory.

Poland's Syndrome is much commoner in males than in females, and usually affects the right side of the body. This data suggest that whatever is happening is not completely random, but just why it happens is not yet understood. The rarity of the condition makes it difficult to study. Most doctors have never encountered a case, and many are unaware of its existence. My own pediatrician brushed it off, saying "No one is symmetrical."

Can it be corrected?

Some cases of Poland's syndrome are more severe than others. The pectoralis major muscle on one side of the body is always at least partially missing, and the hand and fingers on the same side are smaller. Sometimes the arm is only partially developed, or the fingers may be webbed. In females, the breast and nipple may be missing. Plastic surgery—to separate fingers or to implant muscle tissue, for example—can correct many of these features.

Are there complications?

Because the chest wall is lacking muscles, respiratory problems may be commoner. Chest muscles enable a person to cough and thus rid the lungs of bacteria and irritants. Some women may have difficulty nursing a child. Most of the problems with Poland's syndrome are not physical, however. Self-consciousness about one's appearance, and learning to live with the reactions of others, well-intentioned and not, are the greatest challenges for an individual with Poland's syndrome.

My Story—Conclusion

I am fortunate to live very close to a Shriner's Orthopedic Hospital. I took my son to see a geneticist there, who confirmed the diagnosis my neighbor had made. She assured us our son would be fine, and would almost certainly not pass on the condition, should he one day have children of his own. The geneticist predicted that in late adolescence he would become self-conscious about his condition, and she invited us to follow up with them at that point.

How right she was! Amir went from cheerfully informing nearly everyone, "I've got a missing muscle!" in toddlerhood, to refusing to take off his shirt in gym class by high school. He began to avoid any activities that would build up his upper-body muscles and exacerbate the difference between his left and right side. He began insisting that "something be done" about his condition.

So back we went to the Shriner's. There he spoke with a physical therapist who himself had a chest abnormality (although not Poland's Syndrome). Amir talked eagerly to him about his feelings. A surgeon then examined Amir and described in detail the implant procedure which could be done. After weighing the pros and cons, Amir decided that purely cosmetic surgery was not worth the trouble.

No one at the hospital minimized his feelings in any way, or told him he should be lucky his condition wasn't worse. He was taken very seriously, and he was offered choices. He chose to live with his asymmetrical body. Although outwardly no different than he ever was, Amir now feels 1000% better about himself. He's gone back to working out with weights and playing basketball. He's just a normal teen.

Resources:

The National Center for Biotechnology Information, established by the National Institutes for Health, has information on Poland's syndrome here:

<http://www.ncbi.nlm.nih.gov/entrez/dispmim.cgi?id=173800>

<http://www.polands-syndrome.com/> is an extensive website started and maintained by an individual with Poland's Syndrome. It contains a glossary, a fetal development calendar, stories, pictures, links, and much more.

http://www.acay.com.au/~cjr/pol_list.html

is an informative website maintained by an Australian nurse. It has links to a very active online support community for Poland's Syndrome, and other good information for families about disabilities.

The Shriner's Hospitals for Children are equipped to treat Poland's syndrome and other orthopedic problems. There is no charge for their services. Call 1-800-237-5055 or visit http://www.shrinershq.org/Hospitals/Hospitals_for_Children/

My Sources:

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