Dwarfism Overview

By Linda Nicholson, Member of the LPA Medical Advisory Board

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There's been a lot of discussion over the years about the proper way to refer to a child with dwarfism. Many people who have the condition prefer the term "little person" or "person of short stature." For some, "dwarf" is acceptable. For most, "midget" definitely is not. But here's an idea everyone can agree on: Why not simply call a person with dwarfism by his or her name?

Being of short stature is only one of the characteristics that make a little person who he or she is. If you're the parent or loved one of a little person, you know this to be true. But here are some facts that other people may not realize about dwarfism and those who have it.

Dwarfism:

- is a condition characterized by short stature. Technically, that means an adult height of 4 feet 10 inches or under, according to the advocacy group Little People of America (LPA).

- can be caused by any one of more than 200 conditions, most of which are genetic. The most common type, accounting for 70% of all cases of short stature, is called achondroplasia.

- can and most often does occur in families where both parents are of average height. In fact, 85% of children with achondroplasia are born to average-size parents.

Dwarfism isn't:

- an intellectual disability. A person who has dwarfism is typically of normal intelligence.

- a disease that requires a "cure." Most people with the condition can live long, fulfilling lives.

- a reason to assume someone is incapable. Little people go to school, go to work, marry, and raise children, just like their average-size peers.

What Causes Short Stature?

More than 200 conditions are known to cause short stature in a child. Most are caused by a spontaneous genetic mutation in the egg or sperm cells prior to conception. Other conditions are caused by genes inherited from one or both parents. In either of these cases, two average-size parents can have a child with short stature (though this is far more likely to occur with a spontaneous mutation). Similarly, depending on the type of condition causing the short stature, it is possible for little people to have an average-size child.

What prompts a gene to mutate is not yet clearly understood. The change is seemingly random and unpreventable, and can occur in any pregnancy. Generally, when average-size parents have a child with short stature due to a spontaneous mutation, it is rare to have a second child who is also of short stature. However, if parents have some form of dwarfism themselves, the odds are much greater that their children will have it as well. A genetic counselor can help determine the likelihood of passing on the condition in these cases.

Dwarfism has other causes, including metabolic or hormonal disorders in infancy or childhood. Chromosomal abnormalities, pituitary gland disorders (which influence growth and metabolism), absorptive problems (when the body can't absorb nutrients adequately), and kidney disease can all lead to short stature if a child fails to grow at a normal rate.

Types of Short Stature

Most types of dwarfism are known as skeletal dysplasias, which are conditions of abnormal bone growth. They're divided into two types: short-trunk and short-limb dysplasias. People with short-trunk dysplasia have a shortened trunk with longer limbs, whereas those with short-limb dysplasia have an average-sized trunk but small arms and legs.

By far, the most common skeletal dysplasia is achondroplasia, a short-limb dysplasia that occurs in about 1 of every 26,000 to 40,000 babies of all races and ethnicities. It can be caused by a spontaneous mutation in one gene or a child can inherit the gene from a parent who has achondroplasia. People with achondroplasia have a relatively long trunk and shortened upper parts of their arms and legs. They may share other features as well, such as a large head with a prominent forehead, a flattened bridge of the nose, shortened hands and fingers, and reduced muscle tone. The average adult height for someone with achondroplasia is about 4 feet.

Diastrophic dysplasia is another, less common form of short-limb dwarfism. It occurs in about 1 in 100,000 births, and is also sometimes characterized by cleft palate, clubfeet, and ears with a cauliflower appearance. People who have it tend to have shortened forearms and calves (this is known as mesomelic shortening).

Spondyloepiphyseal dysplasias (SED) refers to a group of short-trunk skeletal conditions that affect about 1 in 95,000 babies. Along with achondroplasia and diastrophic dysplasia, it is one of the most common forms of dwarfism. In some forms, a lack of growth in the trunk area may not become apparent until the child is between 5 and 10 years old; other forms are apparent at birth. Often, kids with this disorder also have clubfeet, cleft palate, and a barrel-chested appearance.

In general, dwarfism caused by skeletal dysplasias results in what is known as disproportionate short stature — meaning the limbs are short in comparison with the rest of the body. Metabolic or hormonal disorders typically cause proportionate dwarfism, meaning a person's arms, legs, and trunk are all shortened but remain in proportion to overall body size.

Diagnosis

Some types of dwarfism can be identified through prenatal testing if a doctor suspects a particular condition and tests for it. But most cases are not identified until after the child is born. In those instances, the doctor makes a diagnosis based on the child's appearance, failure to grow, and X-rays of the bones. Depending on the type of dwarfism the child has, diagnosis often can be made almost immediately after birth.

Once a diagnosis is made, there is no "treatment" for most of the conditions that lead to short stature. Hormonal or metabolic problems may be treated with hormone injections or special diets to spark a child's growth, but skeletal dysplasias cannot be "cured." People with these types of dwarfism can, however, get medical care for some of the health complications that are associated with short stature. Problems associated with the different forms of dwarfism involve other body systems — such as vision or hearing — and require careful monitoring.

Possible Complications and Treatments

Short stature is the one quality all people with dwarfism have in common. After that, each of the many conditions that cause dwarfism has its own set of characteristics and possible complications. Fortunately, many of these complications are treatable, so that people of short stature can lead healthy, active lives.

For example, some babies with achondroplasia may experience hydrocephalus (excess fluid around the brain). They may also have a greater risk of developing apnea — a temporary stop in breathing during sleep — because of abnormally small or misshapen airways or, more likely, because of airway obstruction by the adenoids or the tonsils. Occasionally, a part of the brain or spinal cord is compressed. With close monitoring by doctors, however, these potentially serious problems can be detected early and surgically corrected.

As a child with dwarfism grows, other issues may also become apparent, including:

- delayed development of some motor skills, such as sitting up and walking
- a greater susceptibility to ear infections and hearing loss
- breathing problems caused by small chests
- weight problems
- curvature of the spine (scoliosis)
- bowed legs
- trouble with joint flexibility and early arthritis
- lower back pain or leg numbness
- crowding of teeth in the jaw

Proper medical care can alleviate many of these problems. For example, surgery can often bring relief from the pain of joints that wear out under the stress of bearing weight differently with limited flexibility. Surgery also can be used to improve some of the leg, hip, and spine problems people with short stature sometimes face.

Nonsurgical options may help, too — for instance, excessive weight can worsen many orthopedic problems, so a nutritionist might help develop a healthy plan for shedding extra pounds. And doctors or physical therapists can recommend ways to increase physical activity without putting extra stress on the bones and joints.

Helping Your Child

Although types of dwarfism, and their severity and complications, vary from person to person, in general a child's life span is not affected by the dwarfism. Though the Americans with Disabilities Act protects the rights of people with dwarfism, many members of the short-statured community don't feel that they have a disability.

You can help your child with dwarfism lead the best life possible by building his or her sense of independence and self-esteem right from the start. Here are some tips to keep in mind:

- Treat your child according to his or her age, not size. If you expect a 6-year-old to clean up his or her room, don't make an exception simply because your child is small.

- Adapt to your child's limitations. Something as simple as a light switch extender can give a short-statured child a sense of independence around the house.

- Present your child's condition — both to your child and to others — as a difference rather than a hindrance. Your attitude and expectations can have a significant influence on your child's self-esteem.

- Learn to deal with people's reactions, whether it's simple curiosity or outright ignorance, without anger. Address questions or comments as directly as possible, then take a moment to point out something

special about your child. If your child is with you, this approach shows that you notice all the other qualities that make him or her unique. It will also help prepare your child for dealing with these situations when you're not there.

- If your child is teased at school, don't overlook it. Talk to teachers and administrators to make sure your child is getting the support he or she needs.

- Encourage your child to find a hobby or activity to enjoy. If sports aren't going to be your child's forte, then maybe music, art, computers, writing, or photography will be.

- Finally, get involved with support associations like the Little People of America. Getting to know other people with dwarfism — both as peers and mentors — can show your child just how much he or she can achieve.

Reviewed by: Linda Nicholson, MS, MC
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