Idiopathic Hemihypertrophy

OVERVIEW
Your child has been diagnosed with idiopathic hemihypertrophy, which refers to overgrowth of one side of the body. A person with hemihypertrophy will be found to have enlargement on the affected side, including one side of the face and trunk appearing more full and the arm and leg on the same side being longer and larger in diameter. Typically, the percentage difference is small and stays constant with growth and does not change after maturity. Hemihypertrophy in young children is important because there is a risk of certain abdominal cancers and it is important to have regular screening ultrasound exams. The size difference can be a mild cosmetic issue, but it is usually subtle. If the leg length difference is more than 1 inch, treatment with a shoe lift or a growth slowing operation roughly 2-3 years prior to maturity is usually beneficial.

BACKGROUND
The cause of hemihypertrophy is unknown and is the reason for the designation as "idiopathic". Hemihypertrophy also occurs as part of several recognized clinical syndromes which have other visible cutaneous or vascular anomalies, including Proteus syndrome, Klippel-Trenaunay syndrome, neurofibromatosis, and Beckwith-Wiedemann syndrome. Hemihypertrophy without evidence of these other syndromes is also called "non-syndromic" hemihypertrophy. Everyone has a small degree of side to side differences in their face, trunk, and limbs. Roughly 30% of the population will have a detectable small leg length difference, which does not cause any difficulty. However, in hemihypertrophy there is a clear difference in the girth and length of the arm and/or leg. Side to side differences in the face and trunk are often present as well, but are often subtle. The distinction between normal side to side variation and hypertrophy is not clearly defined.

CLINICAL PRESENTATION AND DIAGNOSIS
Hemihypertrophy is believed to be present from birth, but because the infant is small, the side to side difference is also small. As the child grows, the small difference becomes visible. It is often recognized by non-family members, presumably because family members become accustomed to the difference. There are no symptoms or complaints. The leg length difference is usually mild and not a limitation or factor affecting gait or activities. The diagnosis of hemihypertrophy is made by careful physical examination to identify the difference and to rule out
signs of syndromic hemihypertrophy. There are no tests to specifically diagnosis hemihypertrophy. Measurements will be made on physical examination and with special x-rays to quantify the leg length difference.

TREATMENT
There is no specific treatment for hemihypertrophy. Children continue to grow and develop, typically without problems. The appearance of the side to side difference can be a cosmetic issue for some, but often the difference is subtle and most people will not notice the difference unless they look closely. Unfortunately, there are not good options to alter this side to side difference. If the child notices or is old enough for other children to notice, it may help their confidence and acceptance to point out differences in your own face or in faces of other family members.

Leg length discrepancy is managed with the same treatments used for other causes of leg length discrepancy. For hemihypertrophy, usually the difference is small and most of the time no treatment is needed. 30% of all people has a leg length difference up to 1.5 cm and has no difficulty because of it. Fortunately, a small difference is a child, often stays a small difference at maturity. It is believed that the percentage difference from one to the other remains constant. In other words, a toddler with one leg that is 4% longer, would be expect to grow and continue to have one leg be 4% longer. A 5 year old with legs than measure 30 cm and 31.2 cm has a 1.2 cm (4%) difference and would be estimated at maturity to have legs that measure 60 cm and 62.4 cm with a 2.4 cm (4%) difference. In general, small differences less than 2 cm need no treatment at all. Differences between 2-4 cm are usually managed with a shoe lift to partially correct the difference. For differences between 2-6 cm, a growth slowing operation can be done on the long leg roughly 2-3 years prior to maturity to allow the short let to "catch up" and get the legs more equal in length. Regular follow-up visits with the pediatric orthopedic surgeon are important to monitor the difference and to make the best estimate of timing for the growth slowing operation. Leg length differences greater than 5 cm are uncommon in hemihypertrophy, but if present can be managed with surgery to shorten the long leg, or lengthen the short leg. These surgeries are more complicated than the growth slowing operation.

One important issue, is that there is a small, but higher than normal risk for abdominal cancers. Standard recommendations are to get a screening abdominal ultrasound scan every 6 months, until age 6-8 years of age. Detection of these tumors at an early stage significantly improves treatment results. Non-syndromic
hemihypertrophy has been most commonly associated with Wilms' tumor, but also adrenal carcinoma, and hepatoblastoma. However, the majority of patients with Wilm's tumor do not have hemihypertrophy, and no large prospective study of the incidence of tumors in non-syndromic hemihypertrophy has been reported. The presence of Wilm's and other tumors can be identified with abdominal sonography. Clear data for justification and timing of screening is lacking, but on the conservative end of this argument, the standard recommendation is for an abdominal ultrasound every four to six months until the age of eight.

MORE INFORMATION
Further information can be obtained on the internet. Your local public library can help you explore these sources if you are interested. Two good sites for expert and peer reviewed information are the American Academy of Orthopedic Surgeons at www.aaos.org and www.emedicine.com.

FEEDBACK
If you have questions or comments, please contact the office or submit them to the web site at www.pedortho.com.