Evaluation of Leg Length Discrepancy in Children

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Target Audience

This CME activity is intended for physicians, medical students and nurse practitioners. Pediatric emergency department physicians, emergency physicians, pediatricians, and family practitioners will find this information especially useful.

Learning Objectives

After completion of this article, the reader will be able to:

1. Describe what leg length discrepancies are.
2. Summarize common congenital and acquired causes of leg length discrepancy.
3. Discuss important medical and functional problems associated with leg length discrepancies.
4. State the most reliable methods for measuring leg length discrepancy both clinically and radiographically.
5. Discuss treatment options once the leg length discrepancy has been estimated.

Abstract

The authors list common congenital and acquired causes of leg length discrepancy (LLD) to assist the primary care physician with differential diagnosis. Important medical problems associated with LLD are discussed, emphasizing the risk of malignant neoplasm with Beckwith-Wiedemann hemihypertrophy. Potential functional problems caused by LLD in children are discussed. Orthopaedic referral is recommended for LLD predicted to exceed 2 cm. The most reliable methods for measuring LLD clinically and radiographically are presented. Options for orthopaedic management and prediction of LLD are briefly outlined. Int Pediatr. 2004;19(3):134-142.

Key words: Leg length discrepancy (LLD), lower-limb length inequality, hemihypertrophy, growth prediction, epiphysiodesis

Introduction

Approximately 15% of the adult population has a leg length discrepancy (LLD) measuring greater than 1 cm. Most LLDs < 2 cm are idiopathic, due to normal anatomic variation (asymmetry) of the human body. Such minor discrepancies are generally believed to cause few functional problems and often go unnoticed by patients. Larger discrepancies typically have demonstrable underlying causes and often require treatment. The authors discuss a systematic approach to the assessment of LLD, with emphasis on optimal measurement techniques and on avoidance of measurement error.

Presentation of LLD

LLDs exceeding 2 cm may alter normal gait patterns, resulting in a painless limp. Most young children compensate by tiptoe gait on the short limb. Older children may “vault” over the longer extremity, causing an excessive rise in the pelvis with each step. These compensatory mechanisms may result in slight increased energy expenditure, but healthy children with LLD rarely complain of easy fatigability.

Some studies regarding adults suggest that LLDs > 2 cm can result in low back pain, structural scoliosis, and hip degeneration, but another body of literature...
refutes these findings.\textsuperscript{9,11} Certainly, children rarely complain of back or hip pain as a result of LLD. Most often children are evaluated for LLD because the parents suspect a limp or because pelvic tilt is noted during the Adam’s screening test for scoliosis.

### Causes of LLD

While it is not feasible to list all potential causes of LLD, some of the more common etiologies of congenital and acquired LLD are listed in Table 1.

#### Table 1 - Common Etiologies of Leg Length Discrepancy

<table>
<thead>
<tr>
<th>Congenital</th>
<th>Clinical findings:</th>
</tr>
</thead>
<tbody>
<tr>
<td>Proximal focal femoral deficiency</td>
<td>+ Allis, thigh skin dimple</td>
</tr>
<tr>
<td>Developmental coxa vara</td>
<td>+ Allis, limited abduction</td>
</tr>
<tr>
<td>Developmental hip dysplasia</td>
<td>+ Allis, limited abduction</td>
</tr>
<tr>
<td>Fibular hemimelia</td>
<td>Absent lateral toes, valgus ankle</td>
</tr>
<tr>
<td>Tibial hemimelia</td>
<td>Severe varus ankle, leg skin dimple</td>
</tr>
<tr>
<td>Congenital postero medial bowing tibia</td>
<td>Apparent ankle dorsiflexion</td>
</tr>
<tr>
<td>Congenital tibial pseudarthrosis</td>
<td>Neurofibromatosis, anterolateral bowing</td>
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<tr>
<td>Nonsyndromic hemihypertrophy</td>
<td></td>
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</tbody>
</table>

**Physeal growth disturbance**

- Ischemic physeal arrest (Perthes, post-infectious, limb ischemia, septic shock)
- Blount’s disease (tibia vara)
- Radiation therapy
- Juxta-physeal tumor or bone cyst
- Multiple exostosis / osteochondromatosis

**Trauma**

- Traumatic physeal growth arrest
- Fracture malunion (overriding)
- Slipped capital femoral epiphysis (SCFE)

**Hyperemia**

- Posttraumatic overgrowth (common after femur shaft fracture)
- Chronic knee synovitis with overgrowth
  - Chronic osteomyelitis
  - Hemophilia
  - Rheumatoid arthritis
  - Osteoid osteoma
- Arterio-venous malformation (AVM) or hemangiomatosis
- Post-surgical hyperemia

**Neuromuscular**

- Poliomyelitis
- Spastic hemiplegia (cerebral palsy, stroke)
- Spinal cord anomaly (tethered cord, syrinx)

**Syndromes**

- Ollier’s
- Russell-Silver
- Beckwith-Wiedemann
- Proteus
- Klippel-Trenaunay
- Neurofibromatosis
- Conradi-Hunerman
- Vivid cutis marmorata
- Hemiatrophy

<table>
<thead>
<tr>
<th>Clinical findings:</th>
</tr>
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<tbody>
<tr>
<td>Multiple enchondromatosis</td>
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<tr>
<td>Limb asymmetry, growth retardation, clinodactyly</td>
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<tr>
<td>Neonatal hypoglycemia, hemihypertrophy</td>
</tr>
<tr>
<td>Nevi, hemihypertrophy, macrodactyly, hamartoma</td>
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<tr>
<td>Hemangiomata, venous insufficiency</td>
</tr>
<tr>
<td>Local gigantism, macrodactyly, cafe-au-lait nevi</td>
</tr>
<tr>
<td>Stippled epiphyses, short stature</td>
</tr>
<tr>
<td>Marbled skin markings, hemiatrophy</td>
</tr>
<tr>
<td>Facial and corporal hemiatrophy</td>
</tr>
</tbody>
</table>
Many of the congenital anomalies involve hypoplasia or hemihypertrophy of a limb and have telltale skin markings. An anterior skin dimple portends an underlying congenital hypoplasia or hemimelia within the limb segment (Fig. 1). Lower extremity hemihypertrophies and hemiatrophies may also result in unequal limb circumference and foot sizes. All congenital bowing deformities of the tibia result in growth retardation. Infrequently, LLD results from hereditary conditions such as familial exostosis or skeletal dysplasia, so family history is important. The physician should search for history of infection, trauma, or birth complications which may explain LLD. Medications, intraosseous infusions, and growth hormone are not known to cause or aggravate LLD.

Chronic hyperemia may result in limb overgrowth, especially if the hyperemia is near major knee growth plates, so evaluation for asymmetrical pulses, vascular markings, and bruits is prudent. Juxta-physeseal osteonecrosis, as in Perthes disease, radiation therapy, or limb ischemia may cause premature physeseal closure. Hemiatrophy usually results from neurological causes, such as spastic hemiparesis or tethered cord, so examination of the back, feet, and reflexes is important. In summary, the list of possible causes of LLD is extensive, but the differential diagnosis can quickly be narrowed with a thorough history and physical examination.

**Hemihypertrophy**

The term *hemihypertrophy* is defined as enlargement of one side of the body to a greater degree than can be attributed to normal variation. Classic total hemihypertrophy implies enlargement of the face, extremities, trunk, and internal organs on one side. Limited hemihypertrophy may involve a portion of the body on one side. Hemihypertrophy may also be classified as syndromic or non-syndromic.

The primary care physician must carefully assess neonates for hemihypertrophy since it can be associated with severe neonatal hypoglycemia or with malignant tumors, as seen in Beckwith-Wiedemann syndrome (BWS). This syndrome should be considered in infants with hemihypertrophy who are large for gestational age. BWS consists of exophthalmos, macroglossia, gigantism, visceromegaly, abdominal wall defects, and neonatal hypoglycemia. Developmental delay may occur if hypoglycemia is not treated promptly, and BWS has a 7–10% risk of associated malignant tumors, usually prior to age 5.

Hemihypertrophy occurs in only 13% of BWS cases; however, infants with BWS and hemihypertrophy have a 40% chance of developing malignant tumors, the most common being Wilms’ tumor, adrenal cortical carcinoma, or hepatoblastoma. For this reason, tumor screening with thorough clinical examination plus abdominal ultrasound is recommended every 3–4 months until age 7, then every six months until maturity. Annual urinalysis and serum alpha-fetoprotein are also recommended. Genetic evaluation is reasonable to verify the diagnosis and to provide genetic counseling.

Various benign and malignant tumors may occur in other types of syndromic hemihypertrophy, such as neurofibromatosis, Klippel-Trenaunay syndrome, and Proteus syndrome. Non-syndromic
hemihypertrophy lacks vascular or cutaneous lesions, is typically not familial, and has no increased risk of neoplasia.16

**Clinical Assessment LLD**

While medical students are frequently taught to use a tape measure from the malleoli to the umbilicus (or to the anterior superior iliac spines), such landmarks will often result in imprecise estimates of LLD.10,29 For patients who can stand in anatomic position, a more reliable clinical measurement consists of simultaneous palpation of both iliac crests.29 Wood blocks (or a portion of a phone book) can be placed beneath the foot of the shorter limb until the pelvis is leveled by palpation (Fig. 2). A leveled pelvis may be verified by scoliometer using the Adam’s forward bending test. Palpation of the iliac wings may be less reliable in obese patients or in children with significant trunk asymmetry caused by lumbar scoliosis.

If a LLD is suspected by pelvic tilt during standing, the location of discrepancy may be verified by performing the Allis test and the reverse Allis test. The Allis test (also called Galeazzi test) is performed in the supine patient by noting relative knee heights when both hips and knees are flexed 90°.30 This will determine how much discrepancy is located in the thigh segment. The patient is then turned prone with the knees and ankles at 90° (and both hips in neutral rotation) to determine how much LLD is present below the knees. These clinical tests cannot be performed if there exist significant contractures, which limit hip, knee, or ankle motion. When properly used in combination, the standing palpation method and Allis tests can provide a reasonable clinical estimate of LLD.

The clinician can sometimes erroneously diagnose a true LLD when the patient in fact has a functional discrepancy resulting from an angular deformity of one limb or a joint contracture in the lower extremity (LE). For instance, a lower extremity may have symmetrical bone lengths, but will seem too short if there exists a unilateral knee flexion contracture, bowleg, or hip adduction contracture. A lower limb may seem too long if there is an Achilles contracture elevating the heel. Therefore, clinical exam for LLD must include assessment of LE joint motion, angulation, and motor function.

**Radiographic Measurement of LLD**

There are several methods for radiographic measurement of LLD, and each may be optimal in certain circumstances. One of the simplest techniques is to obtain a standing (or supine) AP radiograph of both LEs on a long x-ray cassette. Magnification error is minimal for small children, but the technician must be certain that both knees are extended during the exposure. A standing x-ray will show all sources of discrepancy from the pelvis to the heel, and can show frontal-plane angular deformities such as genu varum.

The scanogram (orthoroentgenogram) is taken with the supine patient lying with a ruler between the lower extremities. Three separate exposures are taken, each perpendicular to the hips, knees, and ankles, thereby avoiding magnification error (Fig. 3). This method is
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readily available and is accurate for older children, but may be less reliable for young children who are likely to move between exposures. Scanograms are inappropriate for angular deformities such as knee flexion contracture or severe genu valgum/varum. In addition, the scanogram does not allow measurement of hindfoot discrepancies as may occur with subtalar coalition or some congenital foot deformities. The scanogram is the most frequently used technique for routine measurement of LLD.

For patients who have significant angular deformities or flexion contractures of the lower extremities, a biplanar CT scanogram is the preferred method for radiographic measurement of LLD\(^1\).\(^2\) The CT scanogram (scout view) has less radiation exposure than a standard scanogram, but is more expensive and often requires another appointment. Unless inclusion of hindfoot length is specifically requested during a CT scanogram, the radiologist will typically provide a measurement from the top of the femoral head to the ankle joint.

**Prediction of LLD**

Prediction of LLD can be difficult because discrepancies may be static, regressive, or progressive.\(^3\)

In order to plan treatment, the orthopaedic surgeon must try to predict how much LLD an untreated patient will likely develop by skeletal maturity. Such predictions facilitate timing of growth-retarding procedures such as epiphysiodesis, and help to determine if limb length equalization is even feasible, given surgical limitations. Because children mature at different ages, most of the techniques for prediction of LLD are based upon skeletal age rather than chronologic age. Skeletal age is typically determined by comparing an AP radiograph of the child's left hand and wrist with radiographs in the Greulich-Pyle atlas.\(^4\)

Skeletal ages are less accurate prior to age 6, but accuracy is improved by placing more emphasis on the hand bones rather than on the carpal bones.\(^5\) Skeletal age should be interpreted with caution if there is hemihypertrophy or hemiatrophy involving the left hand. Use of a normal right-hand radiograph is reasonable if anomalies preclude use of the left hand.

While other methods have been described, the authors will briefly summarize four of the LLD prediction methods in common usage. The arithmetic method\(^6\) provides a rough estimate of growth potential for children older than 10 years. This method assumes that the distal femoral physis grows 10 mm per year and the proximal tibial physis grows 6 mm per year. The method also assumes that boys reach maturity at chronologic age 16, and girls at age 14.

The Green-Anderson growth-remaining charts\(^7\) may be used to estimate growth potential in the distal femoral and proximal tibial physes at various skeletal ages. There are separate charts for boys and for girls. This method has withstood the test of time, and is especially useful if a treatment decision needs to be made without the benefit of serial measurements.

The Moseley graph\(^8\) is a logarithmic representation of the Green-Anderson chart, which allows the growth of both lower limbs to be plotted as straight lines. This method requires at least 2 scanogram measurements, but becomes more accurate if one has the luxury of multiple measurements over many years.
Finally, the multiplier method of Paley et al.\(^3^9\) allows one to estimate future LLD for many congenital anomalies at an early age by assuming that lower extremity proportions will remain constant throughout growth. Each prediction method described above may be appropriate for certain circumstances, and recent studies have shown little difference in accuracy among the various methods.\(^4^0,4^1\) Skeletal ages are generally considered accurate to ± 1 year, which is sufficient to allow equalization of the lower limbs to within 1.5 cm.\(^4^0,4^2\)

**Treatment Options for LLD**

Once the predicted LLD has been estimated, treatment options may be formulated. Localization of the discrepancy within the femur and/or tibia will help determine where treatment should be rendered, since one goal is to maintain symmetry of limb segments. Treatment may also depend on the predicted height of the patient, with shortening procedures less desirable in patients with shorter stature. Mature height may be predicted by using the patient’s skeletal age to extrapolate his growth curve, which is available on the pediatrician’s height-weight chart. The goal of treatment is to obtain nearly equal limb lengths, preserve patient height, maintain body proportions, and minimize surgical risk to the patient. If a patient has a stiff knee or foot drop, it may be desirable to leave the affected limb 1–2 cm short to allow the foot to clear the floor during ambulation.

Virtually all orthopaedic surgeons agree that LLD < 2 cm should be treated non-surgically, with observation, or with an internal (hidden) heel-lift if the patient has a visible limp. Internal heel-lifts are generally limited to 1 cm thickness. Larger lifts will elevate the heel out of the shoe, make the shoe feel tight, or aggravate contracture of the Achilles tendon.\(^4^3\) Therefore, shoe-lifts > 1 cm must generally be added externally on the shoe sole where they are visible. External lifts greater than 5 cm may become quite heavy and unstable, and are poorly tolerated as permanent treatment of LLD. The orthopaedic surgeon will usually prescribe a lift 1–1.5 cm smaller than the actual discrepancy.

Unless a patient has very short stature, a LLD of 2–5 cm is usually treated with shortening of the longer limb. If there is sufficient growth remaining, the longer limb may be safely and gradually shortened with surgical arrest (epiphysiodesis) of the distal femoral and/or proximal tibial growth plates. In recent years, percutaneous epiphysiodesis techniques (Fig. 4) have been developed to minimize scarring.\(^4^4,4^7\) If growth plates are closed, acute femoral shortening of up to 5 cm, may be performed utilizing intramedullary rod stabilization to allow early rehabilitation.\(^4^8\) Tibial shortening is rarely performed due to higher risk of persistent muscle weakness, nonunion, and compartment syndrome.\(^4^9\)

Predicted LLDs of 5–20 cm usually require one or more limb lengthenings, adding contralateral limb shortening for more severe LLD.\(^5^0\) Limb-lengthening techniques involve osteotomy followed by gradual (1 mm per day) distraction with an external fixator. Bone tissue spontaneously regenerates within the gap produced (Fig. 5). The time required for lengthening by “distraction histogenesis” averages 1-2 months per
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Lengthenings greater than 20% of the original bone length are considered very high risk, but lengthenings of 4–5 cm are usually manageable with relatively lower risk of joint subluxation, neurovascular injury, or permanent muscle contracture.

Limb lengthening is a prolonged and somewhat painful undertaking, and may place a psychological burden on the patient and family. The safety and efficacy of limb lengthening has improved significantly since the Ilizarov technique was introduced to North America in the late 1980’s. Even so, with few exceptions, patients with predicted LLD of > 20 cm are typically better candidates for amputation and prosthetic fitting, especially if severe foot deformity is present.

In summary, the primary care physician often has the first opportunity to diagnose LLD and to look for possible congenital, developmental, or genetic causes. The pediatrician must be aware of relatively rare syndromes associated with LLD, especially BWS, which can be associated with neoplasia. Thorough history, physical exam, and plain radiographic imaging will usually reveal the etiology of LLD. Orthopaedic referral is appropriate for management of LLD predicted to exceed 2 cm, but smaller discrepancies rarely require treatment or cause functional problems. The primary care physician and orthopaedic surgeon must work together to assess the patient’s needs and to formulate a treatment strategy which takes into account the medical risks, family support, and access to specialized pediatric orthopaedic care.

References

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Questions

1. Leg length discrepancies measuring less than 2 centimeters at skeletal maturity are usually:
   A. symptomatic
   B. idiopathic
   C. syndromic
   D. congenital
   E. progressive

2. Which of the following correctly describes Beckwith-Wiedemann syndrome?
   A. associated with macroglossia, visceromegaly, and neonatal hyperglycemia
   B. always associated with hemihypertrophy and leg length discrepancy
   C. childhood screening for malignant neoplasia is advisable
   D. associated with intrauterine growth retardation
   E. all of the above

3. Which of the following represents entirely a functional leg length discrepancy?
   A. congenital femoral hypoplasia
   B. congenital fibular hemimelia
   C. congenital posteromedial tibial bowing
   D. unilateral fixed knee flexion contracture
   E. non-syndromic total hemihypertrophy

4. Which of the following radiographic techniques is preferred for measurements of leg length discrepancy in a child with a 40° unilateral knee flexion contracture?
   A. supine AP radiograph of both lower extremities
   B. standing AP radiograph of both lower extremities
   C. scanogram of both lower extremities
   D. standing AP pelvis radiograph
   E. biplanar CT scanogram of both lower extremities

5. A 12-year-old boy with a leg length discrepancy predicted to be 3 centimeters at maturity would typically be managed with:
   A. observation
   B. permanent shoe lift
   C. epiphysiodesis (growth plate arrest)
   D. acute femoral shortening with intramedullary rod fixation
   E. tibial lengthening with distraction histiogenesis and external fixation
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