

## > THE PATIENT

6-month-old girl

# SIGNS & SYMPTOMS

- Leg-length discrepancy
- Asymmetric gluteal folds
- Asymmetric galaxies
  and popliteal fossae
  Positive Galeazzi test

# CASE REPORT

## **NONLINE EXCLUSIVE**

Beth P. Davis, DPT, MBA, FNAP; Amir Barzin, DO, MS; Cristen Page, MD, MPH

Emory University School of Medicine, Department of Rehabilitation Medicine, Division of Physical Therapy, Atlanta, Ga (Dr. Davis); Department of Family Medicine, School of Medicine, University of North Carolina at Chapel Hill (Drs. Barzin and Page)

### bethpdavis@emory.edu

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## **THE CASE**

A healthy 6-month-old girl born via spontaneous vaginal delivery to a 33-year-old mother presented to her family physician (FP) for a routine well-child examination. The mother's prenatal anatomy scan, delivery, and personal and family history were unremarkable. The patient was not firstborn or breech, and there was no family history of hip dysplasia. On prior infant well-child examinations, Ortolani and Barlow maneuvers were negative, and the patient demonstrated spontaneous movement of both legs. There was no evidence of hip dysplasia, lower extremity weakness, musculoskeletal abnormalities, or abnormal skin markings. The patient had normal growth and development (50th percentile for height and weight, average Ages & Stages Questionnaire scores) and no history of infection or trauma.

At the current presentation, the FP noted a leg-length discrepancy while palpating the bony (patellar and malleolar) landmarks of the lower extremities, but the right and left anterior superior iliac spine was symmetrical. The gluteal folds and popliteal fossae were asymmetric, a Galeazzi test was positive, and the right leg measured approximately 2 cm shorter than the left leg. There was no evidence of scoliosis or pelvic abnormalities. Physical examination revealed no ecchymosis or trauma. Orthopedic evaluation by the FP of the hips, knees, and ankles was normal, including negative repeat Ortolani and Barlow maneuvers and normal range of motion. We obtained x-rays of the lower extremities and ordered an orthopedic consultation.

## **THE DIAGNOSIS**

The differential diagnosis included congenital, traumatic, infectious, inflammatory, idiopathic, and neurologic causes.<sup>1-3</sup> The most common etiologies of leg-length discrepancies are summarized in **TABLE 1**.<sup>1-3</sup> Radiographic imaging showed a femur length discrepancy, which was determined to be congenital without indication of trauma or disease; therefore, a diagnosis of congenital femoral bowing was made.

Initial orthopedic evaluation revealed a femur length discrepancy of approximately 2 cm. Plain films showed lateral femoral bowing (FIGURE 1A). Regular interval imaging performed at routine well-child examinations at 19 months, 3 years, and 5 years of age showed progression of the femoral length discrepancy from 2 cm to nearly 5 cm, remaining proportionally constant, as well as increasing genu valgum of the right leg up to 12 degrees (FIGURE 1B-D).

## DISCUSSION

Congenital femoral bowing, which can present as a leg-length discrepancy in infants, is a relatively rare finding with an incidence of 1 per 52,000 births.<sup>4</sup> Our patient presented

### TABLE 1

## Congenital and acquired causes of leg-length discrepancy<sup>1-3</sup>

Congenital causes	Acquired causes
Hemihypertrophy/hemiatrophy	Cancer or other neoplastic changes
Idiopathic conditions	Idiopathic conditions
Klippel-Trenaunay-Weber syndrome	Blount disease
Proteus syndrome	Legg-Calvé-Perthes disease
Limb hypoplasia syndromes	Infection
Distal	Osteomyelitis
Congenital posteromedial bowing	Purpura fulminans
Fibular hemimelia	Septic arthritis
Tibial hemimelia	Inflammation
Proximal	Juvenile rheumatoid arthritis
Congenital short femur	Pigmented villonodular synovitis
• Coxa vara	Neurologic
Developmental dysplasia of the hip	Cerebral palsy
Femoral hypoplasia	Closed head injury
Proximal femoral focal deficiency	Myelomeningocele
Skeletal dysplasias	Peripheral nerve injury
Chondrodysplasia punctate	Polio
Fibrous dysplasia	Spinal cord injury or tumor
Multiple hereditary exostoses	Trauma
Neurofibromatosis	Acute bone loss
Ollier disease	Burns
Synovial osteochondromatosis	Fracture (Salter-Harris, slipped capital femoral epiphysis)
	latrogenic
	Irradiation

with an isolated limb deformity, but congenital femoral bowing is recognized as a clinical feature of several skeletal dysplasias (TABLE 2).<sup>5</sup>

### What's recommended

The American Academy of Pediatrics recommends routine age-appropriate physical examination without specifying leg-length assessment.<sup>6</sup> There is insufficient evidence, according to the US Preventive Services Task Force and the American Academy of Pediatrics, regarding the value of routine infant hip and leg-length assessment for developmental dysplasia of the hip and other musculoskeletal abnormalities; however, both agree that abnormal findings require follow-up and management.<sup>6-8</sup>

## Congenital femoral bowing requires plain film diagnosis

Following physical examination, diagnosis of congenital femoral bowing should be confirmed by plain films. Plain radiography remains the main imaging modality for proximal focal femoral deficiency and fibular hemimelia, and appropriate identification of the osseous abnormalities seen on radiographs allows for accurate classification of congenital femoral bowing, prognosis, and surgical planning. (Minor malformations associated with congenital leg-length discrep-

Congenital femoral bowing may present clinically as a leg-length discrepancy with no indication of trauma or disease.

## FIGURE 1 Plain films reveal increasing femoral length discrepancy



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Initial plain films at 6 months of age revealed an approximately 2-cm femur length discrepancy and lateral femoral bowing (A). Regular interval imaging performed at 19 months (B), 3 years (C), and 5 years of age showed progression of the femoral length discrepancy to nearly 5 cm with up to 12 degrees of genu valgum of the right leg (D).

ancies are not typically identified as being part of a larger syndromic diagnosis.<sup>4</sup>) The patient should subsequently be referred to an orthopedist for monitoring and to establish a long-term management plan.

### Early diagnosis can improve treatment outcome

Both early diagnosis of congenital femoral bowing and prediction of leg-length discrepancy at skeletal maturity can influence potential treatment options, which range from conservative management (eg, watchful waiting, physical therapy, shoe lifts, orthotics, bracing) to surgical intervention. Several models have been used to predict skeletal growth, including the Moseley straight line graph, Green and Anderson growth curve, Amstutz method, and Paley's multiplier method.4,9-14

Intervention for leg-length discrepancy generally is dictated by the magnitude of the inequality and the presence of functional deficits and/or pain.<sup>2</sup> If the degree of femur angulation begins to affect structural development, surgical intervention should be con-

sidered to align development and/or correct the discrepancy. Physical therapy, shoe lifts, orthotics, and bracing are treatment options for managing smaller discrepancies.<sup>2,15</sup>

Our patient. The physician (CP) reviewed treatment options with the family that included watchful waiting, use of a shoe lift and/or orthotics, and bracing. The family chose watchful waiting due to the structural integrity of the patient's other major joints and her relatively preserved function. The patient demonstrated mild gross motor delay at routine well-child visits at 9, 12, and 15 months, but was walking by 19 months. Her development was otherwise normal as documented via regular developmental screenings at routine well-child examinations. The patient participated in home physical therapy to maintain strength, flexibility, and functional mobility, and she was able to ambulate by walking on her right toes.

Surgery. Ultimately the patient underwent medial distal femoral hemiepiphysiodesis of the right lower extremity at 6 years of age due to increasing leg-length discrepancy and lateralization of the patella from the val-

## TABLE 2 Skeletal dysplasias associated with angulated femurs<sup>5</sup>

Angulated femur is sometimes present in:

Thanatophoric dysplasia (types I and II)

Osteogenesis imperfecta (types II and III)

Short-rib thoracic dysplasia with or without polydactyly (types I–IV; asphyxiating thoracic dysplasia, Ellis–van Creveld syndrome)

Hypophosphatasia (infantile, perinatal lethal)

Femoral hypoplasia (femoral facial syndrome, focal femoral hypoplasia)

Type II collagen defects

- Achondrogenesis type 2/hypochondrogenesis (severe form)
- Hypochondrogenesis
- Kniest dysplasia
- Platyspondylic lethal skeletal dysplasia, Torrance type
- Spondyloepiphyseal dysplasia congenita

Angulated femur is always present in:

Antley-Bixler syndrome

Campomelic dysplasia

Cartilage hair hypoplasia (McKusick type)

Cumming syndrome

Kyphomelic dysplasia

Prenatal infantile cortical hyperosteosis

Stüve-Wiedemann syndrome

gus deformity. The patient's mother reported that she did well postoperatively, with increased range of motion, improved physical capabilities, and reduced discomfort in the right leg. She continued to participate in physical therapy and had routine follow-up with her physical therapist, her FP, and orthopedist.

■ A second surgery. At approximately 8 years and 9 months of age, the orthopedist noted that the patient's leg-length discrepancy had increased, and she had right extensor mechanism malalignment and severe patellar subluxation. The patient subsequently underwent surgery to remove the existing hardware, including right extensor mechanism realignment via a Roux-Goldthwait procedure (with reconstruction of the medial patellofemoral ligament and anterior cruciate ligament), as well as left distal femoral epiphysiodesis. She did very well postoperatively and continues to participate in physical therapy approximately once weekly. She has had an improvement in her gait and stability using shoe lifts.

## THE TAKEAWAY

A routine well-child examination can be an opportunity to identify congenital musculoskeletal problems. Congenital femoral bowing is a relatively rare finding<sup>4</sup> that may present as a leg-length discrepancy. With proper evaluation, including visual inspection, palpation, range-of-motion testing, and special tests as needed (eg, Galeazzi test, Ortolani and Barlow maneuvers), early intervention is possible if a leg-length discrepancy is noted. Close monitoring of gait abnormalities at routine well-child visits is essential.

Physical therapy, shoe lift therapy, and surgical approaches are treatment options for leg-length discrepancy,<sup>2</sup> and early intervention can improve treatment outcomes.<sup>14</sup> Understanding how to manage congenital femoral bowing over time is important in providing options and counselling patients and their families.<sup>15</sup>

Treatment of leg-length discrepancy in pediatric patients requires long-term management with a team approach that includes patients and their families. The goal of intervention is to reduce physical and emotional trauma, while addressing complications and maintaining function of the affected limb, as well as the whole body.<sup>15</sup> JFP

#### CORRESPONDENCE

Beth P. Davis, DPT, MBA, FNAP, Emory University School of Medicine, Department of Rehabilitation Medicine, Division of Physical Therapy, 1462 Clifton Road NE, Suite 312, Atlanta, GA 30342; bethpdavis@emory.edu.

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