

CASE REPORT

ONLINE EXCLUSIVE

> THE PATIENT
1-day-old neonate

- > SIGNS & SYMPTOMS
- Hyperextension of the bilateral knees
 - No knee fractures or dislocation on x-ray
 - Normal prenatal ultrasounds and lab screenings

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> THE CASE

A 29-year-old G7P2315 woman gave birth to a girl at 37 weeks via spontaneous vaginal delivery. APGAR scores were 9 and 9. Birth weight was 2760 g. Cardiovascular and pulmonary examinations were normal (heart rate, 154 beats/min; respiratory rate, 52 breaths/min). Following delivery, the neonate appeared healthy, had a lusty cry, and had no visible craniofacial or cutaneous abnormalities; however, the bilateral knees were hyperextended to 90° to 110° (FIGURE 1A).

The mother had started prenatal care at 7 weeks with 10 total visits to her family physician (JD) throughout the pregnancy. Routine laboratory screening and prenatal ultrasounds (including an anatomy scan) were normal. She had a history of 3 preterm deliveries at 35 weeks, 36 weeks, and 36 weeks, respectively, and had been on progesterone shots once weekly starting at 18 weeks during the current pregnancy. She had no history of infections or recent travel. Her family history was remarkable for a sister who gave birth to a child with thrombocytopenia absent radius syndrome.

THE DIAGNOSIS

The neonate tolerated passive flexion of the knees to a neutral position. Hip examination demonstrated appropriate range of movement with negative Ortolani and Barlow tests. The infant's feet aligned correctly, with toes in the front and heels in the back, and an x-ray of the bilateral knees showed no fractures or dislocation.

Based on the clinical examination and x-ray findings, we made a diagnosis of congenital genu recurvatum. A pediatric orthopedics consultation was obtained, and the knees were placed in short leg splints in comfortable flexion to neutral on Day 1 of life. She was discharged the next day.

FIGURE 1A

Our patient at ... birth



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Both of the patient's knees were hyperextended to 90° to 110°.

DISCUSSION

Congenital genu recurvatum, also known as congenital dislocation of the knee, is a rare condition involving abnormal hyperextension of the unilateral or bilateral knees with limited flexion.¹ Reports in the literature are limited, but there seems to be a female predominance among known cases of congenital genu recurvatum.² The clinical presentation varies. Finding may be isolated to the knee(s) but also can present in association with other congenital abnormalities, such as developmental dysplasia of the hip, clubfoot, and hindfoot and forefoot deformities.^{3,4}

Diagnosis is made clinically with radiographic imaging

Diagnosis of congenital genu recurvatum is made clinically and can be confirmed via radiographic imaging of the knees.⁵ Clinical diagnosis requires assessment of the degree of hyperextension and palpation of the femoral condyles, which become more prominent as the severity of the hyperextension increases.⁶ X-rays help assess if a true dislocation or subluxation of the tibia on the femur has oc-

curred. Based on the clinical and radiographic findings, congenital genu recurvatum typically is classified according to 3 levels of severity: grade 1 classification only involves hyperextension of the knees without dislocation or subluxation, grade 2 involves the same characteristic hyperextension along with anterior subluxation of the tibia on the femur, and grade 3 includes hyperextension with true dislocation of the tibia on the femur.¹ Grades 1 and 2 on this spectrum technically are diagnosed as congenital genu recurvatum while grade 3 is diagnosed as a congenital dislocation of the knee,⁷ although the 2 terms are used interchangeably in the literature. We classified our case as a grade 1 congenital genu recurvatum based on the clinical and radiographic findings.

Congenital knee hyperextension has intrinsic and extrinsic causes

Hyperextension of the knees at birth may be caused by various intrinsic or extrinsic factors. Intrinsic causes may include breech position, lack of intrauterine space, trauma to the mother, quadriceps contracture or fi-

...1 week of life



Long leg splints were placed with a maximal flexion of 20° to 30°.

... 3 weeks of life



The patient had improved knee flexion at 3 weeks, and the long leg splints were removed.

➤ **Clinical diagnosis includes palpation of the femoral condyles, which become more prominent as the severity of the hyperextension increases.**

brosis, absence of the suprapatellar pouch, deficient or hypoplastic anterior cruciate ligament, pathological tissues, arthrogryposis, or genetic disorders such as Larsen syndrome or achondroplasia.⁶

Extrinsic causes may include traumatic dislocation during the birthing process³ or intrauterine pressure leading to malposition of the joints. When intrauterine pressure is combined with reduced intrauterine space, this phenomenon is known as packaging disorder.⁶ Entanglement of the umbilical cord around the legs of the fetus during development may be another potential factor.¹ Of note: Cases involving both extrinsic or intrinsic etiologies can present with associated abnormalities that include congenital dislocation of the hip, congenital hip dysplasia, spina bifida, and/or cleft palate—in addition to knee hyperextension.

■ **The exact etiology in our patient** was unknown, but we determined the cause was extrinsic based on the lack of other genetic abnormalities. We initially considered a possible connection between our patient's diagnosis and her family history of thrombocytopenia absent radius syndrome, but it was later determined that both were isolated cases and the limb abnormalities were coincidental.

■ **Treatment options and outcomes** for extrinsic and intrinsic etiologies depend on the severity of the hyperextension and any associated abnormalities, as well as the time in which therapy is initiated.¹ Reduction of the hyperextension within 24 hours of birth has been associated with excellent outcomes.⁸ Regardless of the cause, all cases of congenital genu recurvatum should first be treated conservatively. Evidence has suggested that conservative therapy involving early gentle manipulation of the knee combined with serial splinting and casting should be the first line of treatment.⁶ If initial treatment attempts fail or in cases occurring later in life, surgical interventions (eg, quadriceps release procedures such as percutaneous quadriceps recession or V-Y quadriceps-plasty, proximal tibial closing-wedge, anterior displacement osteotomy) likely is warranted.^{6,9}

■ **Our patient.** At 1 week of life, our patient's short leg splints were replaced with long leg splints with a maximal flexion of 20° to 30° (FIGURE 1B). Weekly follow-ups with serial casting were initiated in the pediatric orthopedics clinic. At 3 weeks of life, the patient's knee flexion had improved and the splints were removed (FIGURE 1C). Upon clinical examination, the bilateral knees were extended to a neutral position, and both could be actively and passively flexed to 90°. The patient was referred to Physical Therapy to perform range of movement exercises on the knees.

At 8 weeks of life, the bilateral legs were in full extension, and knee flexion was up to 130°. Physical therapy for knee range of movement exercise was continued on a weekly basis until 6 months of life, then twice monthly until the patient was 1 year old. Ultimately, the hyperextension was corrected, and the patient started walking at around 16 months of age. Her prognosis is good, and she will be able to participate in low-impact sports, after consulting with her orthopedist.

THE TAKEAWAY

Congenital genu recurvatum is a rare condition that presents with abnormal hyperextension of the knee(s) with limited flexion. Early diagnosis and assessment of the severity of the hyperextension is crucial in determining the type of intervention to pursue. Conservative management entails serial casting and splinting to increase knee flexion. If conservative management fails or if the diagnosis is made later in life, surgical options often are pursued. **JFP**

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