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The Novel Use of the Lateral Scanogram to Detect Cruciate Deficiency in Children With Congenital Femoral Deficiency

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Abstract

Congenital femoral deficiency (CFD) is often associated with cruciate ligament deficiency. The lateral scanogram may be a potential solution to some limitations for detecting instability associated with cruciate ligament deficiency. This qualitative case study identified two children with congenital femoral deficiency who were assessed with a lateral scanogram and had their results correlated to the clinical examination and MRI. Both cases identified a child with congenital femoral deficiency, one with a total leg length discrepancy (LLD) of 12 cm and the next with 6.5 cm. The weight-bearing lateral scanogram revealed anterior tibial translation, indicating knee instability. Both patients will undergo anterior cruciate ligament (ACL) reconstruction prior to limb lengthening. The lateral scanogram is a useful imaging modality that is capable of detecting anterior tibial translation, and thereby knee instability, in children with congenital femoral deficiency. Larger studies utilizing and evaluating the benefits of lateral scanograms are warranted.

Categories: Pediatric Surgery, Radiology, Orthopedics

Keywords: cruciate deficiency, anterior tibial translation, proximal femoral focal deficiency, congenital femoral deficiency, lateral scanogram

Introduction

Congenital femoral deficiency (CFD), a spectrum of pathology that encompasses congenital short femur and proximal femoral focal deficiency (PFFD), is a rare congenital anomaly with an incidence of 1.1-2.0 in 100,000 live births worldwide [1-4] and a male-to-female ratio of 1:2 [2]. A single underlying genetic cause for CFD has not been elucidated, and most available data points to the teratogenic or idiopathic somatic mutations of cells within the developing limb bud versus a hereditary etiology [5,6].

A deficiency of the cruciate ligaments is commonly associated with CFD and other various congenital longitudinal deficiencies of the lower limb [7-10]. A systematic review of CFD and associated ligament deficiencies in the knee noted that there is not enough support in the literature to back the routine reconstruction of the cruciate ligaments in patients with CFD [4]. In higher-functioning individuals, congenital absence is often well tolerated [10]. However, in children undergoing limb lengthening with severe CFD, anteroposterior knee subluxation occurs frequently and may often be symptomatic [4,10-12].

Despite the association between CFD and cruciate ligament deficiency, routine screening and treatment for these patients vary among practice, especially in patients undergoing limb lengthenings [4,7,11,13]. While anterior scanograms, or 2-3 radiographs of the lower extremities in the anteroposterior plane stitched together, are used to assess leg length discrepancy (LLD), the use of a scanogram from the lateral view has not yet been described as a method for screening in CFD patients suspected to have cruciate ligament deficiencies. Our case report describes two patients with CFD that were found to be cruciate ligament-deficient on physical examination and by lateral scanogram. Our goal is to describe how lateral scanograms may be used to effectively assess instability associated with cruciate ligament deficiency in patients with

Case Presentation

Case 1

A 10-year-old female with a history of right congenital femoral deficiency and valgus deformity presented for limb lengthening (Figure 1). She currently wears a shoe lift of 5.5 cm and complains of pain after walking and occasional instability in her knee.

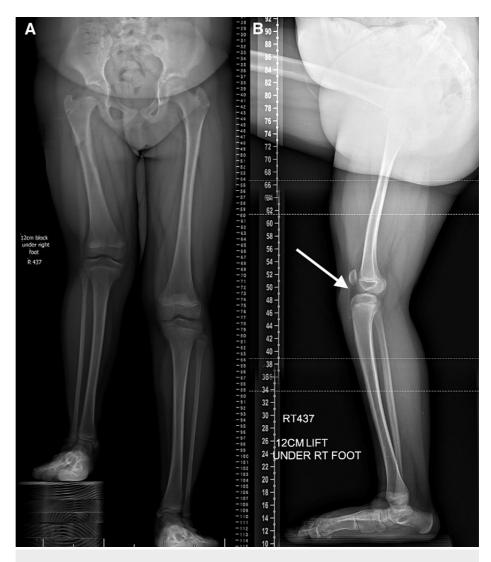


FIGURE 1: Anterior and Lateral Scanograms of Patient 1

(A) Anterior scanogram depicting 12 cm leg length difference of the right leg. (B) Lateral scanogram showing anterior tibial translation (white arrow)

On physical examination, the patient walked with a short-legged gait with genu valgum. With a 12 cm block under her right foot, her pelvis was level and spine straight on Adam's forward bend test. She had no end point on the anterior drawer test.

Anterior scanogram revealed a right femoral length of 28.6 cm versus left femoral length of 40.2 cm and right tibia length of 31.2 cm versus left tibia length of 31.8 cm. The lateral scanogram revealed an anteriorly translated tibia with a posterior tibial slope of 15 degrees. A follow-up MRI revealed anterior cruciate ligament (ACL) deficiency and a posterior cruciate ligament (PCL) with decreased ligament caliber (Figure 2).

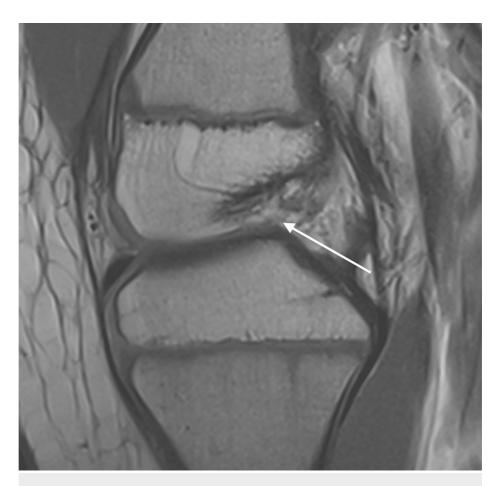


FIGURE 2: Lateral Right Knee MRI of Patient 1

White arrow: the absence of anterior cruciate ligament

After discussion with family, the patient was scheduled to undergo a combined intra-articular and extra-articular ACL reconstruction and extra-articular PCL reconstruction with the ipsilateral iliotibial band, right distal medial femoral hemi-epiphysiodesis to address her knee valgus, and contralateral epiphysiodesis. The plan to address her leg length discrepancy is two femoral lengthenings with internal lengthening nail.

Case 2

A six-year-old male presented with his father for LLD (Figure 3). He had a history of right clubfoot with only four toes that was treated with serial casting and tenotomy. His LLD has been managed with orthotics and currently has an external lift on the right shoe.



FIGURE 3: Anterior and Lateral Scanograms of Patient 2

(A) Anterior scanogram depicting leg length discrepancy of 6.5 cm (red arrow). (B) Lateral scanogram showing anterior tibial translation (white arrow) indicating cruciate ligament deficiency

On physical examination, he ambulated unassisted and had wide symmetric hip abduction, right genu valgum, and positive Galeazzi with the right femur 1.5 cm shorter than the left. His right tibia appeared 5 cm shorter than the left. His right foot only had four rays but otherwise normal dorsiflexion and plantarflexion. His pelvis was level with a 6.5 cm block under his right foot. His right knee demonstrated no end point on anterior drawer.

Anterior scanogram revealed a right femoral length of 36.1 cm versus left femoral length of 37.2 cm and right tibia length of 26.8 cm versus left tibia length of 31.2 cm. His lateral scanogram revealed the anterior translation of the tibia with the knee in extension. A follow-up MRI revealed the absence of the ACL and PCL (Figure 4).

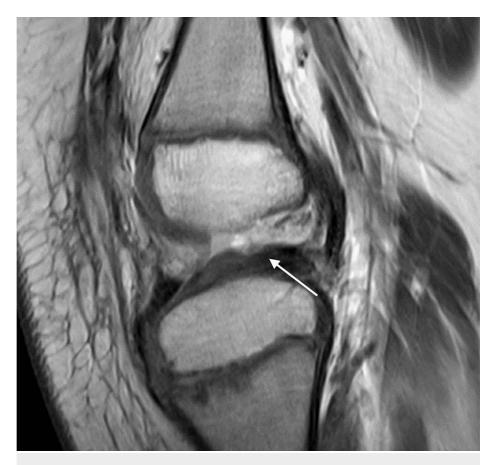


FIGURE 4: Lateral View of Right Knee MRI of Patient 2

Note the absence of the anterior cruciate ligament (white arrow) and anterior tibial translation

This patient was diagnosed with right fibular hemimelia, right CFD, right genu valgum, ACL and PCL deficiency with knee subluxation, and an LLD of 6.5 cm (the projected LLD was 9.3 cm). The patient underwent hemi-epiphysiodesis of the medial distal femur and extra-articular and intra-articular reconstruction of the ACL and extra-articular PCL construction in preparation for limb-lengthening surgery. Leg length discrepancy will be addressed with the future lengthening of his tibia and possibly femur.

Discussion

Cruciate ligament deficiency in patients with CFD undergoing lengthening is important to identify for surgical planning. Recurrent swelling and instability have previously been reported in patients with congenital aplasia of the ACL [9,14], and subluxation or dislocation during lengthenings also may occur in patients with CFD [9,11,15]. Torzilli et al. described in cadaveric models how the knees with a sectioned ACL displayed a "substantial anterior neutral-position shift" of the tibia during loading compared to native knees [16]. This was corroborated by Beynnon et al. who radiographed living knees with torn ACLs versus healthy knees, with and without loading, and found a fourfold increase in anterior translation [17]. They posed this increased laxity in the sagittal axis as a potential risk for meniscal and osteochondral damage associated with cruciate deficiency. Furthermore, the risk of severe early knee arthritis may be increased during the limb-lengthening process in patients with CFD and preoperative knee instability, due to abnormal pressure on articular cartilage, resulting in degeneration [18]. This was corroborated by Jeong et al., who reported an arthritis rate of 78% in CFD patients after lengthenings and a trend toward a positive correlation between preoperative knee instability and degenerative arthritis at the last follow-up [11].

Due to these potential consequences, children with CFD are routinely evaluated with anterior and lateral scanograms in our practice. After the anterior scanogram is obtained, the lateral view can be taken accordingly (Figure 5). The patient is radiographed at a source image receptor distance of 102 cm. The patient should stand upright facing laterally such that their index limb is positioned distal to the tube and their femoral condyles parallel to the radiograph's beam. The handlebars of the scanogram apparatus should be adjusted to chest height for the patient to grab for stability, and the anterior-most hip should then be flexed to 90 degrees. Depending on habitus or strength, this limb may need to be stabilized by a stool or radiology technician to prevent leaning too much on the index limb.



FIGURE 5: Proper Positioning for Lateral Scanogram

(A) The scanogram apparatus that the patients stand in for anterior and lateral scanograms. (B) Proper positioning for a lateral scanogram. Note that the anterior-most hip (right leg) is flexed to 90 degrees and may be supported at the knee. Although an adult is used for modeling in this figure, the proper positioning of all patients regardless of age, BMI, or gender should remain consistent with what is displayed. Image credits: X Author

The majority of ACL deficiencies can be diagnosed through history and clinical examination. The anterior drawer, Lachman, and pivot shift test are commonly used to detect clinically the absence or tear of an ACL. For surgical planning, the confirmation of knee instability, and the detection of concomitant bony abnormalities, correlation to imaging is necessary. The lateral scanogram is helpful to obtain compared to standard radiographs of the knee because it allows for the visualization of the entire lower limb in two planes while weight-bearing. Standard lateral radiographs provide the visualization of the bony anatomy from an orthogonal view to the anterior scanogram, but lateral scanograms may be quickly obtained after the anterior scanogram while also providing views of the proximal femur and distal tibia, which may be the pathological areas of interest during treatment. In contrast to routine screening with MRIs, which can cost on average 3,300 USD [19], the weight-bearing aspect of the lateral scanogram provides insight to how the knee behaves during their gait, and the distinct anterior translation of the tibia provides evidence of cruciate deficiency and instability. Furthermore, many patients with CFD require routine screening for their LLD by anterior scanogram; thus, a rotation in stance for the lateral scanogram is a prudent and efficient method for detecting concomitant instability.

Conclusions

CFD is commonly associated with cruciate ligament deficiency that can result in an unstable knee. Efficient screening for cruciate deficiencies in these patients is important to ensure proper care and prevent potential adverse outcomes of any planned lengthening surgeries. We reported two cases of patients with CFD who were found to have a cruciate deficiency on lateral scanogram by anterior tibial translation. To our knowledge, this is the only study that reports on the use of lateral scanograms in CFD patients. Lateral scanograms are an effective method to evaluate for bone or ligamentous deformity/deficiency in patients with CFD.

Additional Information

Disclosures

Human subjects: Consent was obtained or waived by all participants in this study. Baylor College of Medicine issued approval H-52441. H-52441: Leg Length Discrepancy in Children and Adolescents: Retrospective Analysis Approval valid from 09/21/2022 to 09/06/2027 Dear Dr. Hill, The Institutional Review Board for Human Subject Research for Baylor College of Medicine and Affiliated Hospitals (BCM IRB) is pleased to inform you that the research protocol named above was reviewed and approved by expedited

procedures on 09/21/2022 by board 2. The study does not require continuing review but will require a 5 year renewal check in with the IRB office. You will receive an email renewal reminder notice prior to study expiration; however, it is your responsibility to assure that this study is not conducted beyond the expiration date. Please be aware that only IRB-approved informed consent forms may be used when written informed consent is required. Any changes in study or informed consent procedure must be submitted to the IRB as an amendment for review and approval prior to implementation unless the change is necessary for the safety of subjects. In addition, you must inform the IRB of adverse events encountered during the study or of any new and significant information that may impact a research participants' safety or willingness to continue in your study. Research that has been approved by the BCM IRB may be subject to further appropriate review and approval or disapproval by officials of the institution(s) where the research will be conducted. However, those institutional officials may not approve the research if it has not yet been approved by the IRB. The BCM IRB is organized, operates, and is registered with the United States Office for Human Research Protections according to the regulations codified in the United States Code of Federal Regulations at 45 CFR 46 and 21 CFR 56. The BCM IRB operates under the BCM Federal Wide Assurance No. 00000286, as well as those of hospitals and institutions affiliated with the college. Sincerely yours, Julie Katkin, MD, BA Institutional Review Board for Baylor College of Medicine and Affiliated Hospitals . Conflicts of interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following: Payment/services info: All authors have declared that no financial support was received from any organization for the submitted work. Financial relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work. Other relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

References

- Paley D, Chong DY, Prince DE: Congenital Femoral Deficiency reconstruction and lengthening surgery.
 Pediatric lower limb deformities. Sabharwal S (ed): Springer, Cham, Switzerland; 2016. 361-425.
- Ifesanya A, Omololu B: Case report: proximal focal femoral deficiency in Ibadan a developing country's perspective and a review of the literature. Nig J Paed. 2012, 39:75-8. 10.4314/njp.v39i2.8
- Doğer E, Köpük SY, Cakıroğlu Y, Cakır O, Yücesoy G: Unilateral isolated proximal femoral focal deficiency.
 Case Rep Obstet Gynecol. 2013, 2013:637904. 10.1155/2013/637904
- Leite CB, Grangeiro PM, Munhoz DU, Giglio PN, Camanho GL, Gobbi RG: The knee in congenital femoral deficiency and its implication in limb lengthening: a systematic review. EFORT Open Rev. 2021, 6:565-71. 10.1302/2058-5241.6.200075
- Hamanishi C: Congenital short femur. Clinical, genetic and epidemiological comparison of the naturally occurring condition with that caused by thalidomide. J Bone Joint Surg Br. 1980, 62:307-20. 10.1302/0301-620X.62B3.7410462
- Paley D, Shannon CE, Nogueira M, Chiari C, Harris M: Can adding BMP2 improve outcomes in patients undergoing the SUPERhip procedure?. Children (Basel). 2021, 8:495. 10.3390/children8060495
- Chomiak J, Podškubka A, Dungl P, Ošt'ádal M, Frydrychová M: Cruciate ligaments in proximal femoral focal deficiency: arthroscopic assessment. J Pediatr Orthop. 2012, 32:21-8. 10.1097/BPO.0b013e31823d34db
- 8. Johansson E, Aparisi T: Missing cruciate ligament in congenital short femur . J Bone Joint Surg. 1983, 65:1109-15.
- Kaelin A, Hulin PH, Carlioz H: Congenital aplasia of the cruciate ligaments. A report of six cases. J Bone Joint Surg Br. 1986, 68:827-8. 10.1302/0301-620X.68B5.3782255
- Saldanha KA, Blakey CM, Broadley P, Fernandes JA: Defining patho-anatomy of the knee in congenital longitudinal lower limb deficiencies. J Limb Lengthen Reconstr. 2016. 2:48-54. 10.4103/2455-3719.182576
- Jeong C, Inan M, Riddle EC, Gabos PG, Bowen JR: Knee arthritis in congenital short femur after Wagner lengthening. Clin Orthop Relat Res. 2006, 451:177-81. 10.1097/01.blo.0000229295.44592.30
- Gabos PG, El Rassi G, Pahys J: Knee reconstruction in syndromes with congenital absence of the anterior cruciate ligament. J Pediatr Orthop. 2005, 25:210-4. 10.1097/01.bpo.0000153874.74819.29
- 13. Grill F, Dungl P: Lengthening for congenital short femur. Results of different methods . J Bone Joint Surg Br. $1991, 73:439-47.\ 10.1302/0301-620X.73B3.1670446$
- Mitsuoka T, Horibe S, Hamada M: Osteochondritis dissecans of the medial femoral condyle associated with congenital hypoplasia of the lateral meniscus and anterior cruciate ligament. Arthroscopy. 1998, 14:630-3. 10.1016/S0749-8063(98)70063-5
- Karger C, Guille JT, Bowen JR: Lengthening of congenital lower limb deficiencies. Clin Orthop Relat Res. 1993, 236-45.
- Torzilli PA, Deng X, Warren RF: The effect of joint-compressive load and quadriceps muscle force on knee motion in the intact and anterior cruciate ligament-sectioned knee. Am J Sports Med. 1994, 22:105-12. 10.1177/036354659402200117
- Beynnon BD, Fleming BC, Labovitch R, Parsons B: Chronic anterior cruciate ligament deficiency is associated with increased anterior translation of the tibia during the transition from non-weightbearing to weightbearing. J Orthop Res. 2002, 20:332-7. 10.1016/S0736-0266(01)00115-2
- 8. Eldridge JC, Bell DF: Problems with substantial limb lengthening. Orthop Clin North Am. 1991, 22:625-31.
- Hayatghaibi SE, Alves VV, Ayyala RS, Dillman JR, Trout AT: Transparency and variability in pricing for pediatric outpatient imaging in US children's hospitals. JAMA Netw Open. 2022, 5:e220736. 10.1001/jamanetworkopen.2022.0736