Femoral bifurcation with ipsilateral tibia hemimelia: Early outcome of ablation and prosthetic fitting

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Abstract

Femoral bifurcation and tibia hemimelia are rare anomalies. Hereby, we present a case report of a 2-year-old boy who first presented in our orthopedic clinic as a 12-day-old neonate, with a grossly deformed right lower limb from a combination of complete tibia hemimelia and ipsilateral femoral bifurcation. Excision of femoral exostosis, knee disarticulation and prosthetic fitting gives satisfactory early outcome.

Key words: Early outcome, femoral bifurcation, tibia hemimelia

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Introduction

Bifurcation of the femur and tibial agenesis are rare anomalies.[1-3] Congenital deficiency of the tibia or tibial hemimelia occurs with an incidence of approximately 1 per 1 million live births.[4] This rare lower limb anomaly is characterized by deficiency of the tibia with relatively intact fibula, which was classified by Jones in 1978,[5] Kalamchi and Dawe in 1985,[6] and Webers in 2008.[7]

Tibia hemimelia may be complete or incomplete, unilateral or bilateral, familial or sporadic, and it affects both genders.[7,8]

Tibia hemimelia and femoral bifurcation may occur independently in isolation.[1,9] More commonly, they co-exist and are accompanied by other congenital anomalies of the limbs or other parts of the body.[1,4,10]

Some of the reported associated anomalies include deficiency of the radius or ulna, Gollop-Wolfgang complex, facial cleft, oligodontia, sacral agenesis, proximal focal femoral deficiency, congenital pyloric stenosis, tracheoesophageal fistula, congenital heart disorders, anorectal atresia, hemivertebra, tibia hemimelia-split hand/foot syndrome (TH-SHFM), Mirror (duplication) foot or hand, absent digits, etc.[11-14] The VACTERL association has been reported in tibia aplasia.[15]

Hereby, we report our findings of a Nigerian with complete tibia hemimelia and ipsilateral femoral bifurcation. This combination is extremely rare and there is paucity of reported cases in African children.

Case Report

A 12-day-old male, delivered by a para 4+0, 30-year-old woman at term, presented with deformities in the right lower limb. The pregnancy and labor were uneventful. He had physiologic jaundice which cleared without any intervention. The patient was said to be very active and well, except for the deformity. There was no family history of any congenital deformity and his other three siblings were normal. No history of infection, drug use or alcohol ingestion in the mother during pregnancy was reported and there was no history suggestive of diabetes mellitus in the mother. The parents were both subsistence farmers.

Examination revealed a very active neonate with gross...
discrepancy in the appearance of the right and left lower limbs [Figure 1]. He was not pale, and was afebrile, had physiologic jaundice which cleared within a week of presentation without any intervention.

Head and neck region was essentially normal. Chest was normal in appearance; he had clear lung fields, heart rate was 136 beats/min, and no added sound was present.

Abdomen was full, moved with respiration, and there was no organomegaly. The perineum and external genitalia were normal.

Both the upper limbs were essentially normal. There were areas of hypopigmentation of the skin on the medial aspects of the left thigh and right leg. The left lower limb was grossly normal. The right thigh was hypoplastic compared with the left. There was a prominent bony swelling protruding from the anteromedial side of the distal femur; the overlying soft tissue was mobile.

There was right knee flexion contracture, hypoplastic leg, varus and adduction deformity of the right foot with absence of the right medial malleolus. The patella was not palpable and could not be located with ultrasonography. All toes were present. There was no neurovascular deficit. The spine was essentially normal.

X-ray showed a normal proximal femur, right anteromedial distal femoral exostosis with diaphyseal extension of its sessile base, giving a Y-shaped right femur (femoral bifurcation). The right distal femoral ossification center was initially absent but later appeared in the X-ray taken at 11 months of age [Figure 2]. The X-ray of other bones of the body and ultrasonography of the abdomen did not reveal any anomaly.

A diagnosis of Jones type Ib right tibia hemimelia with ipsilateral femoral bifurcation was made.

Excision of the exostosis, centralization of the fibula, and later on, limb lengthening procedure were initially planned for. Symes amputation was also discussed.

Parents, however, opted for disarticulation and excision of the exostosis to avoid multistage surgery with uncertain prognosis.

The patient had right knee disarticulation and excision of the exostosis at age 13 months. He is presently fitted with a prosthesis with which he has started ambulation.

**Discussion**

Femoral bifurcation is rare and is usually associated with tibia aplasia and other deformities.

The clinical presentation of a patient with femoral bifurcation and tibia hemimelia will depend on the severity of the hemimelia, age at presentation and other associated anomalies. Our patient was quite healthy except for the deformity in the affected right lower limb. His condition was sporadic as there was no family history of any congenital limb anomaly.

The morphology is varied in femoral bifurcation.\(^{[16]}\) In this patient, the main femur was placed laterally, while the bifurcated portion presented like an anteromedial solitary exostosis with a cartilage cap [Figure 2]. The femur was flattened over a wide area as opposed to what is commonly seen in solitary osteochondroma. Unlike that reported by Ogden, the only distal secondary femoral ossification center was laterally placed in the 11\(^{th}\) month X-ray film; this was absent in the 12\(^{th}\) day X-ray film. The
cartilage-capped exostosis causing the Y-appearance of the femur was excised, leaving the main femur with the distal secondary ossification center which was flat in shape and also looked hypoplastic.

The patient had Jones type Ib tibia hemimelia with severe knee flexion deformity and weak quadriceps power and a large disparity in the lengths of the right and left legs.

Tibia hemimelia often produces major limb length discrepancy and foot deformity. Factors to be considered in deciding whether to perform reconstructive or ablative surgery include the severity of the aplasia, the expected leg length discrepancy, the anomalies of the foot, the status of the knee and quadriceps muscle.\(^{15,16,17}\)

The traditional method for treating complete tibia deficiency is knee disarticulation as the outcome of Brown centralization of the fibula was usually less satisfactory, especially with severe knee flexion deformity and weak quadriceps power.\(^{18,19}\)

With the advent of the Ilizarov frame, many more limb salvage procedures are being performed for severe type I tibia hemimelia with functioning quadriceps and foot. After a successful centralization, limb length equalization is another daunting challenge to overcome. Complications following limb lengthening are related to the severity of the discrepancies. The use of Ilizarov technique has improved the outcome of limb salvage procedures in this severe form.\(^{17,20}\)

When there is severe limb length discrepancy, limb lengthening may not succeed and a secondary amputation may be necessary.\(^{20}\) The result of early disarticulation with prosthetic fitting is more certain and gives satisfactory result when performed early as patients usually accept it like congenital amputation.\(^{13,14,19}\)

In our environment, ablative surgeries are not easily accepted, but in this patient, it was accepted fairly easily. This may be due to the severely disfiguring nature of this combination of deformities and the uncertain outcome of reconstructive surgeries. The fact that the parents also pay for the treatment amidst scarce resources might also have influenced the early consent for ablative surgery. In environments where treatments are not funded by patients or parents, economic considerations are less important in giving consent for surgery.

Our patient with severe limb length discrepancy, knee flexion deformity and weak quadriceps had excision of the femoral exostosis, knee disarticulation and prosthetic fitting, and he is now ambulant.

**Conclusion**

We conclude that excision of the bifurcated femur, knee disarticulation and prosthetic fitting give good early outcome in this rare condition of bifurcated femur with ipsilateral complete hemimelia of the tibia.

**References**


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