

Terminal hemimelia of the lower extremity: absent lateral ray and a normal fibula: reply to comments by Dr. Hootnick

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We thank Dr. Hootnick for his comments and his interest in our paper.

The purpose of our paper was to demonstrate the disease entity which had absent lateral rays and a normal fibula as the mildest form of postaxial hypoplasia of the lower extremity. But, Dr. Hootnick asserted that the absent rays are not lateral ones but midline ones and that absent rays are not a clinical feature of postaxial hypoplasia but result from vascular agenesis.

Dr. Hootnick mentioned that Fig. 1a, an AP radiograph in our paper, demonstrates on gross inspection absence of the second and third metatarsals. However, we considered the fourth and fifth metatarsals to be absent in this figure for several reasons. First, the fifth metatarsal base on the normal side, which is an insertion site for the peroneus brevis, is different from the most lateral metatarsal base on the affected side. Second, the lateral two toes of the affected side are not curved in the same manner as the fourth and fifth toes of the normal side. Third, if the midline metatarsal is missing, the first metatarsal and lateral ones should be divergent because of absence of the transverse metatarsal

ligament. Actually, most papers dealing with fibular hemimelia indicated that the absent rays are lateral ones [1–3, 5–7].

Dr. Hootnick [4] demonstrated that there was an association of malformation of bone and arteries in skeletal dysplasia of the lower limb with an absent fibula. It may be that abnormal vascularisation during embryogenesis results in skeletal dysplasia, but proof is certainly lacking for this hypothesis. The hypothesis of abnormal vascularisation cannot explain why just the lateral condyle of the distal femur is involved and the fibula is more commonly involved than the tibia, etc. Actually, vascular dysgenesis could be one manifestation of the whole musculoskeletal dysplasia of postaxial hypoplasia.

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