



UNIVERSITY of
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MEDICAL CENTER

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DEPARTMENT OF IMAGING SCIENCES

Imaging Sciences Interesting Cases

CASE 309

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CLINICAL PRESENTATION: Patient is a 5-year-old female with shoe fitting problem and history of complex reconstruction of both feet at age 11 months for bilateral polydactyly and syndactyly.

IMAGING FINDINGS: Image findings at age 3 months of age (**Fig. 1**) reveal polydactyly of the bilateral first digits, i.e. pre-axial polydactyly. The left first metatarsal is broad and shortened. The left duplicated digit exhibits phalanges, but no duplication of the metatarsal. The right first digit is well developed, while the duplicated right digit exhibits proximal phalangeal and metatarsal shortening.

Image findings at 5 years of age (**Fig. 2**) exhibit interval growth and development, status post-bilateral reconstruction. The first metatarsal bones are shortened and widened compared to normal. The first Lisfranc joint of the right foot is incongruent. There is hyperextension of both first metatarsophalangeal joints. There is hallux varus deformity.



Figure 1: Plain film of the feet at age 3 months.



Figure 2: Plain film of the feet at age 5 years.

DIAGNOSIS: Polydactyly of the feet

Discussion:

1 Chronicles 20:6 King James Version of the Bible

And yet again there was war at Gath, where was a man of great stature, whose fingers and toes were four and twenty, six on each hand, and six on each foot and he also was the son of the giant.

Polydactyly is a congenital abnormality resulting in supernumerary digits that often results in a difficulty of fitting shoes. The extra digit may range from unarticulated vestigial tissue (Type B) to a well-developed functioning digit (Type A). Temtamy and McKusick classified polydactyly into pre-axial, central, and post-axial types. In the hands, the most common side of polydactyly is the ulnar (post-axial), followed by radial (pre-axial), and lastly the interdigital (central). In the feet, the most common side of polydactyly is the lateral (fibular or post-axial) at 60%; ~15% first digit (tibial or pre-axial); ~15% fourth digit (central type) [3]. HOX13 is one gene with an associated genetic mutation. Simple polydactyly is autosomal dominant with incomplete penetrance and expressivity. In one surgical review, polydactyly was the most common congenital abnormality of the feet at 45% [3].

Polydactyly (or hyperdactyly) may be associated within a developmental syndrome or occur alone as an autosomal dominant trait. In the Radiology of Syndromes, Metabolic Disorders, and Skeletal Dysplasias [5], there are 44 listed post-axial polydactyly syndromes and 28 pre-axial syndromes. Polydactyly may occur with or without syndactyly, which is fusion of two digits: the combination is polysyndactyly. Combined hand and foot forms occur in about 1/3 of the cases; these may be ipsilateral or contralateral. The isolated form is more common in blacks versus whites, at approximately 10:1 (IR = 3.6-10.7/1000 in blacks and 0.3-1.6/1000 in whites). It also has a male predominance.

Pre-axial polydactyly is more common in Asian populations, accounting for 90% of cases in South China, Hong Kong, and Japan. Post-axial polydactyly in white children is often syndromic and autosomal recessive.

Diagnosis may be made on clinical exam, x-ray and/or fetal sonogram. On fetal sonogram, fetal finger buds can be seen as early as 9-10 weeks gestation. Associated abnormalities are seen in about 15% of polydactyly children at birth, however this rises to 55% in children with rare forms of polydactyly. Surgical reconstruction is usually performed at about 1 year of age for well-developed duplicated digits, but can be performed earlier for vestigial soft tissue. Radiographs should be obtained pre-operatively in AP and Lateral projections, with weight bearing (age-dependent).

This patient has sexdactyly in bilateral feet, evident on plain radiographs. Follow-up radiographs were utilized to monitor surgical correction and subsequent changes with growth, especially as there have been relevant clinical concerns, i.e. shoe fitting. MR was also utilized in this patient for developing clinical concerns, but not for initial pre-surgical evaluation or primary diagnosis.

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