A healthy 2-year-old girl was referred for toenail thickening and yellow discoloration; she was otherwise asymptomatic. The patient had no history of diabetes, infection, or trauma to her toenails. Physical examination revealed both great toenails to have marked thickening of the nail plate with closely spaced transverse superficial ridging, lack of luster, and dark tan-yellowish discoloration (Figure 1). There was no subungual debris. There was mild separation of the nail plate from the underlying nail bed (onycholysis) and subungual hemorrhage on her right great toenail. Micronized toenail clippings failed to grow dermatophytes or nondermatophyte yeasts or molds.

The bilateral presentation, persistence, absence of a history of trauma, and negative fungal culture confirm the diagnosis of congenital malalignment of the great toenails. This condition, which is often misdiagnosed and treated as onychomycosis, may occur in 1% to 2% of children (personal communication: R. Baran. E-mail, June 24, 2003). Congenital malalignment of the great toenail describes the condition in which the longitudinal axis of the nail is laterally deviated from birth1 (Figure 2). A malaligned nail matrix causes angular lateral nail plate growth. The toenails develop transverse ridges, appear thickened, and acquire gray, green, or brown discoloration.1 The ridges occur at regular intervals and appear to result from repeated microtrauma to the nail matrix. Hemorrhage and infection cause discoloration.2 In addition, they may be tender to palpation. One or both great toenails may be involved, and other toenails are occasionally also affected. Associated paronychia frequently develops.

Properly fitting footwear reduces microtrauma that causes transverse ridging. Treatment may involve surgical realignment of the nail matrix.2 Spontaneous improvement by 5 and 10 years of age, however, has also been reported.3 Although toenail disorders in children are rare, recognition of congenital malalignment of the great toenail can reduce the cost in time, money, and adverse treatment effects spent on children mistakenly treated for onychomycosis.

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