Congenital diastasis of the inferior tibiofibular joint: case report and treatment analysis

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Congenital diastasis of the inferior tibiofibular joint is a rare anomaly, seen at birth. It is commonly associated with a variety of conditions: talipes equinovarus deformity of the foot, an unduly prominent and distally placed medial malleolus, accentuated tibia vara and tibial intorsion, widened inferior tibiofibular joint and tibial shortening. The condition was first reported in 1972 by Tuli and Varma[1] (2 cases), followed by Bose[2] (1976, 1 case), Jones and associates[3] (1978, 4 cases) and Onimus and colleagues[4] (1990, 4 cases). To date, in the English orthopedic literature, 17 children have been reported with congenital tibiofibular diastasis.[1–9]

All children presented clinically with associated talipes equinovarus deformity of the foot of varying severity. Often noted later were the associated aspects of the congenital diastasis, widening of the ankle mortise, tibial intorsion and a short tibia. All had tibial hypoplasia of varying severity, with evident hypoplasia of the lateral distal plafond. The talus was often wedged and elevated between the tibia and fibula. The fibula was frequently hypertrophic and on lateral radiographs was seen crossing the tibia anteriorly.

We report here our experience with 1 child with this unusual anomaly. We also developed and present a treatment algorithm for congenital tibial diastasis based on an analysis of the English literature.

Case report

A 6-day-old girl was referred to the Children's Hospital of Eastern Ontario in Ottawa, a major pediatric referral centre, because of talipes equinovarus deformity of the left foot. The child was the product of a normal 37-week pregnancy. Both the mother (28 years old and healthy) and father (Italian, unrelated) were nonsmokers with no relevant medical history.

Examination of the child revealed a correctable talipes equinovarus deformity with an otherwise anatomically normal left foot, associated with widening of the ankle mortise (Fig. 1). The medial malleolus was unduly prominent and the lateral malleolus appeared distally displaced. The left leg was 1 cm shorter than the right due to tibial hypoplasia. The tibia was internally rotated and bowed anterolaterally. Ankle plantar and dorsiflexion were normal. The other limbs and the spine were clinically normal.

Further investigations revealed an ostium primum atrial septal defect and an atrioventricular septal defect. Recurrent attacks of episodes of aspiration of food led to the diagnosis of gastroesophageal reflux. No chromosomal or renal abnormalities were detected. Radiologic studies of the tibia confirmed the tibial bowing, dislocation of the proximal and distal tibiofibular joints, and displacement of the distal tibia posteriorly and medially relative to the talus. The talus was laterally displaced (Fig. 2).

The child was treated by serial casting at 1-week intervals for 6 weeks, at which time the talipes equinovarus was corrected. She was then fitted with a Dennis Browne brace at the age of 3 months. Three months later, the foot had maintained its correction, and the bowing and the tibial intorsion were markedly improved. At the time of writing, the left leg was 2 cm shorter than the right. More recent radiologic studies revealed a hypoplastic distal tibial epiphysis and metaphysis (Fig. 3). The treatment plan was to maintain the foot in a plantigrade position by bracing with monitoring of the limb-length inequality. The child will require tibial limb-lengthening procedures in the future. No mortise reconstruction was planned because the ankle mortise was not functionally compromised, and she had full plantar and dorsiflexion.

Discussion

Of the 18 cases of congenital tibio- fibular diastasis (including ours) that have been reported in the English literature,[1–9] 11 were boys and 7, girls. In 9, the diastasis

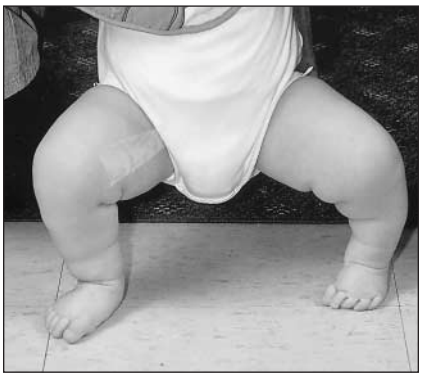

FIG. 1. Clinical photograph of the child with congenital tibiofibular diastasis at 11 months of age. The tibia was 1.8 cm short with marked tibial intorsion.

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was right-sided; in 7, left-sided; and in 2, bilateral (Table 1). One child had a family history of limb malformation. Some patients had associated congenital anomalies in the same leg. Eight had hypoplasia or aplasia of the first ray, and 2 had tarsal coalition. Four children had a concomitant proximal tibiofibular subluxation. One child had a hypoplastic femur. Four had contralateral limb malformations, including distal femoral bifurcation, tibial hypoplasia and hemimelia, and dysplasia of the first ray of the contralateral foot. Four patients also had associated hand abnormalities (lobster claw hand and syndactyly). Only 1 child had lumbosacral vertebral anomalies. Five children were noted to have visceral anomalies, including cardiac, genitourinary and anal defects. Our patient also had an atrial and ventricular septal defect of the heart (Table 1).

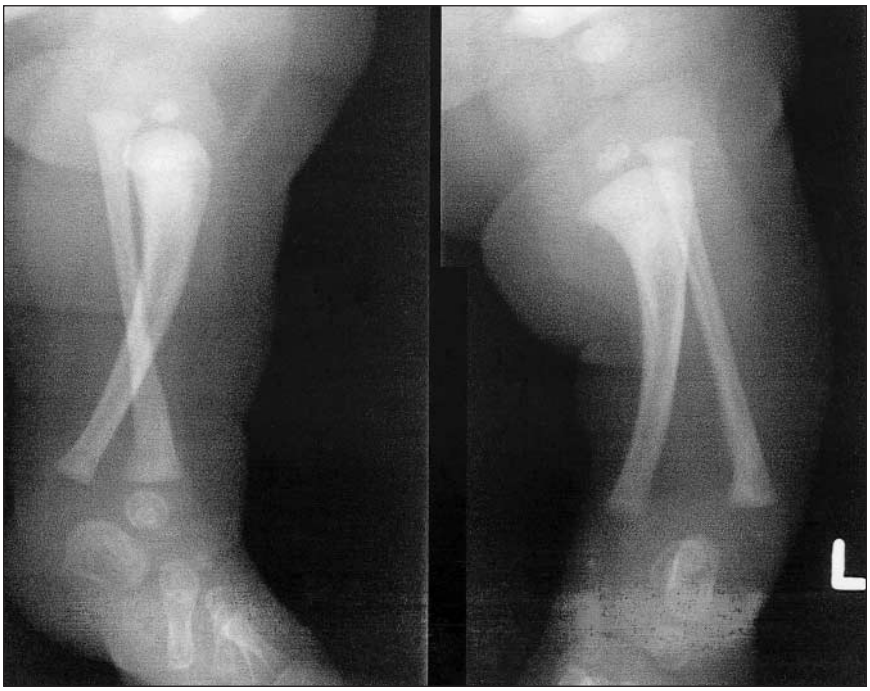

FIG. 2. Initial lateral (left) and anteroposterior radiographs of the child at 6 weeks of age, illustrating the tibiofibular diastasis in both coronal and sagittal planes, tibial bowing into varus and tibial shortening.

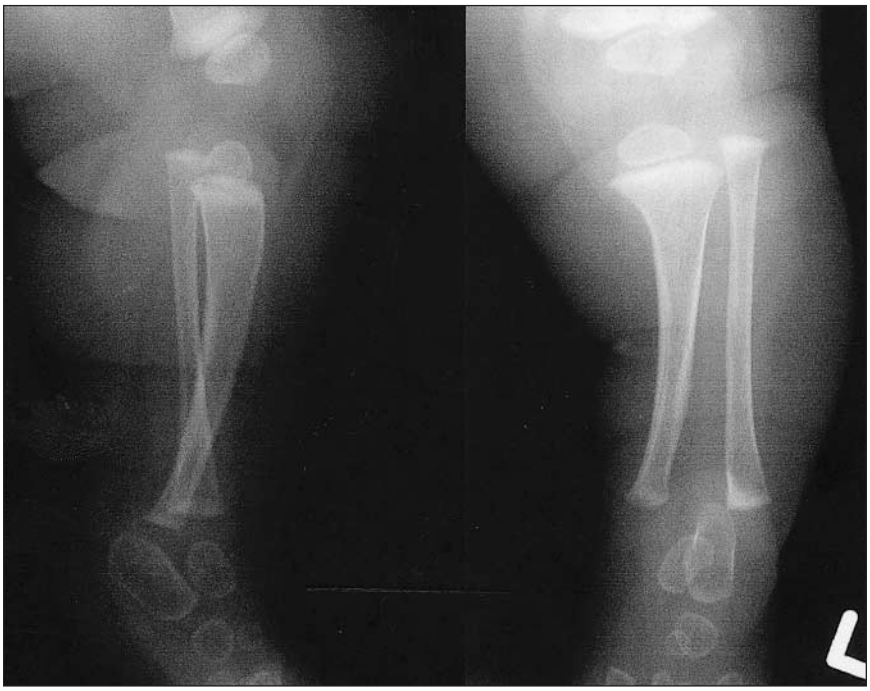

FIG. 3. Lateral (left) and anteroposterior radiographs illustrating a hypoplastic distal tibial epiphysis and metaphysis.

The exact pathogenesis of congenital inferior tibiofibular diastasis is unknown. Tuli and Varma[1] described this condition as an osteochondrosis. Bose[2] thought that congenital inferior tibiofibular diastasis was a form of congenital tibial hypoplasia, an anomaly with a spectrum of clinical and radiographic appearances. This concept was shared by Jones and associates,[3] who classified congenital tibiofibular diastasis as a variant of the congenitally aplastic tibia with short tibia and intact fibula (type IV). Because shortening concerns the tibia only in some cases and both tibia and fibula in most cases reported, a distinction should be made between 2 varieties of congenital diastasis of the tibiofibular joint.[4] In this regard, we have proposed a classification of congenital inferior tibiofibular diastasis to reflect the spectrum of the anomaly (Table 2). In this classification our child had type III tibial diastasis.

Three clinical problems need to be addressed in congenital inferior tibiofibular diastasis: the equinovarus deformity, the ankle diastasis, and the limb-length inequality. Historically, the cornerstone of treatment has been open reduction of the tibiofibular diastasis to reconstitute the ankle mortise.[1,2,7] Several authors attempted this procedure but found it difficult and hazardous, and perhaps unnecessary because the joint is inherently stable regardless of the diastasis.[4] More recent reports have shown good results without reconstruction of the ankle mortise.[3–6,8] Other treatment modalities for the foot deformity have included serial casting, application of braces and posteromedial release, Achilles tendon lengthening, talectomy, ostetomy of the talus and calcaneus, and Syme's amputation.[10–12] On the basis of the treatment experience in the literature[1–9] (Table 1) and our experience, we have formulated a treatment algorithm for the management of this unusual anomaly based on a comprehensive critical review of the cases in the literature and our close 2-year follow-up of the child reported here (Fig. 4). The goal

Table 1

Reported Cases of Congenital Diastasis of the Inferior Tibiofibular Joint*

Series	Sex	Side	Associated anomalies: Ipsilateral	Associated anomalies: Congenital	Treatment of reported cases
Tuli and Varma 1972[1]	F	R	Proximal fibular subluxation; tibial hypoplasia	None	Surgical restoration of ankle mortise; brace
	F	R	Tibial hypoplasia	None	Surgical restoration of ankle mortise; Achilles tendon lengthening; braces
Bose 1976[2]	M	R	Dysplastic 1st ray; proximal fibular subluxation; aplastic talus; tibial hypoplasia	Hypoplastic 1st ray of left foot	Surgical restoration of ankle mortise; Achilles tendon lengthening; medial release; braces
Matthews et al 1977[5]	F	R	Tibial hypoplasia	Left cleft hand; cardiac abnormalities	Serial casting; further surgery planned
Jones et al 1978[3]	M	L	Tibial hypoplasia	Hypoplastic right tibia	Treatment refused
	M	R	Tibial hypoplasia	Left tibial hypoplasia; dysplastic 1st ray of left foot; imperforate anus; hypospadias	None
	M	L	Tibial hypoplasia	None	Posteromedial release; Syme's amputation
	M	R	Tibial hypoplasia	None	Tibia–fibula osteotomy; talectomy; Syme's amputation advised
D'Ambrosio 1979[6]	M	B	Tibial hypoplasia; bilateral absent 1st ray	None	Serial casting; surgery contemplated
Sedgwick and Schoenecker 1982[7]	F	R	Tibial hypoplasia; anterior tibial bow; absent 1st ray	Claw hand on left; lobster claw hand on right	Surgical restoration of ankle mortise; epiphyseal closure of left proximal tibia and distal femur
Wolfgang 1984[9]	M	L	Tibial hypoplasia	R femoral bifurcation; R complete tibial paraxial hemimelia; cleft R hand; absent 3rd ray in L hand and syndactyly of 4th & 5th rays	Serial casting; orthosis; Syme's amputation
Gabarino et al 1985[8]	M	L	Tibial hypoplasia; absent 1st ray, congenital tarsal fusion; hypoplastic femur	Lumbosacral vertebral anomalies	Medial release; Achilles tendon lengthening; osteotomy of talus & calcaneus; epiphyseal closure of right proximal tibia & distal femur; planned right femoral shortening
	M	B	Tibial hypoplasia	Cardiac abnormalities; hypospadias	Serial casts; bilateral posteromedial release; left fibular osteotomy; planned limb-length equalization
Onimus et al 1990[4]	M	R	Proximal tibiofibular subluxation; tibial hypoplasia; aplasia of scaphoid and 1st metatarsal	None	Achilles tendon lengthening; tibial osteotomy; tibial lengthening; femoral lengthening
	F	R	Tibiofibular synostosis; proximal fibular hypoplasia; tibial dysplasia; aplasia of the lateral distal tibia; talar dysplasia	Syndactyly of the 3rd and 4th fingers, both hands; urethrovaginal fistula; bifid tongue	Achilles tendon lengthening; tibial lengthening; tibial osteotomy
	M	L	Aplasia of ceiling of mortise; tibia intorsion; aplasia of 1st ray; talocalcaneal coalition	None	Ilizarov lengthening and derotation; proximal fibular and distal tibial osteotomies
	F	L	Aplasia of ceiling of mortise; aplasia of both lateral and medial rays of the foot	None	Posterior release; casting; splinting; planned leg rotational and lengthening osteotomy
Present report	F	L		Cardiac abnormalities; gastroesophageal reflux disease	Serial casting; Dennis Browne brace; planned limb lengthening

* The presentation in all cases was talipes equinovarus. Superscripted numbers are reference citations. M = male, F = female, R = right, L = left, B = bilateral.

Table 2

Proposed Classification of Children With Congenital Diastasis of the Inferior Tibiofibular Joint

Type	Tibia	Fibula	Diastasis	Foot
I	Hypoplastic; short; intorsion	Normal; longer than tibia; there may be proximal dislocation	Vertical	Supinated
II	Hypoplastic; distal physeal dysplasia	Hypoplastic, same length as tibia	Horizontal	Equinovarus
III	Hypoplastic; short; bowed anterolaterally; intorsion	Hypoplastic, longer than tibia, bowed laterally	Horizontal	Equinovarus; lateral ray hypoplastic or absent

should be a functional plantigrade foot. For severe tibial shortening or a malformed foot that cannot be maintained in a weight-bearing position, a Syme's amputation is still an option[10] but should be deferred until this is an obvious solution. Onimus and colleagues[4] stated that posteromedial release does not always correct the varus deformity because varus does not result from soft-tissue retraction but rather from the skeletal deformity. They also stated that Achilles tendon lengthening and posterior release usually correct the equinus, and other operations may not be necessary. Adduction of the foot can be corrected through a double upper fibular and inferior tibial derotation osteotomy.[4] Internal rotation of the tibia initially can be managed orthotically with a Dennis Browne splint and later, if necessary, by derotation ostetomy or correction with the Ilizarov system during the lengthening procedure. Limb-length inequality, which is the main issue in the older child or children who present late, can be managed by limb-lengthening procedures according to preference. With modern methods of limb lengthening, we would expect good results.

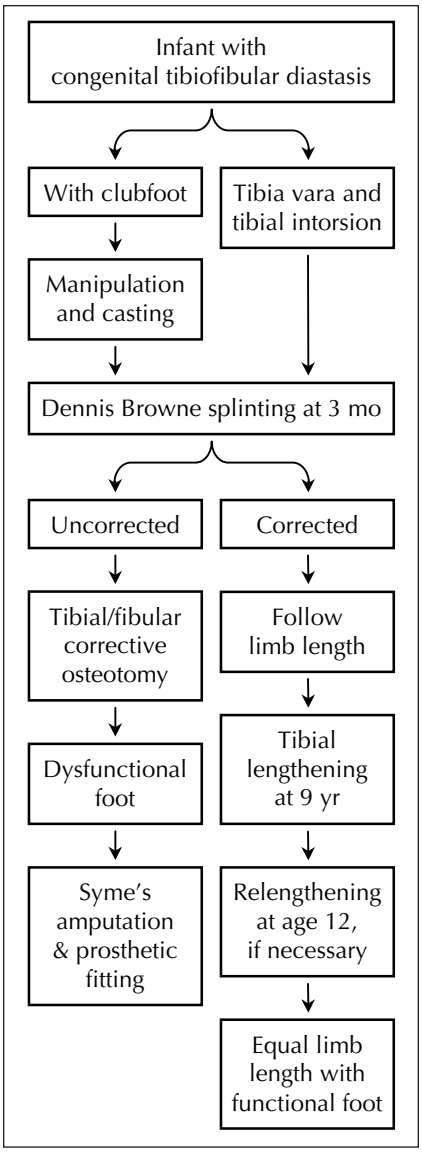

FIG. 4. Treatment algorithm for congenital tibiofibular diastasis.

Conclusions

The child with congenital diastasis of the inferior tibiofibular joint must be investigated for other congenital malformations, which are frequently associated with tibiofibular diastasis. These children have 3 clinical problems: clubfoot, ankle diastasis and limb-length inequality. The ankle diastasis may be compatible with adequate ankle function, and over-treatment of this component of the triad should be avoided. Tibiofibular diastasis is a variant of tibial hypoplasia, and limb-length inequality will be the major deficiency, not the diastasis. We recognize that the child in our report has not been followed up to skeletal maturity; however, based on the comprehensive critical review of similar cases in the literature plus a close 2-year follow-up of this child, we believe that the proposed classification and treatment algorithm, although speculative, will facilitate the treatment plans for children with congenital tibiofibular diastasis.

Competing interests: None declared.

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