Congenital anterolateral tibial bowing associated with polydactyly: case report and literature review

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ABSTRACT
The congenital anterolateral tibial bowing associated with polydactyly (CABTP) is an extremely rare deformity. To the best of our knowledge, only 21 cases have been documented in the English literature. Although there is a strong relationship between anterolateral bowing of the tibia, congenital pseudoarthrosis of the tibia (CPT), and neurofibromatosis type 1 (NF1), CATBP is a specific entity associated with a more favorable prognosis. We report a new CABTP case and our literature review on the previously reported cases of CABTP concerning its associated deformities, diagnosis, and treatment.

Key words: Tibial bowing; polydactyly; hallux.
Level of Evidence: IV

Incurvación anterolateral congénita de la tibia asociada con polidactilia. Reporte de un caso y revisión bibliográfica

RESUMEN
La incurvación anterolateral congénita de la tibia asociada con polidactilia (*congenital anterolateral tibial bowing associated with polydactyly*) es una deformidad extremadamente rara. Solo se han documentado 21 casos en la bibliografía inglesa. Aunque existe una estrecha relación entre la incurvación anterolateral de la tibia, la seudoartrosis congénita de la tibia y la neurofibromatosis tipo 1, la incurvación anterolateral congénita de la tibia asociada con polidactilia es una entidad específica con un pronóstico más favorable. Comunicamos otro caso y también revisamos los casos de este cuadro ya reportados con respecto a las deformidades asociadas, el diagnóstico y el tratamiento.

Palabras clave: Incurvación tibial; polidactilia; hallux.
Nivel de Evidencia: IV

INTRODUCTION

Congenital tibial bowing is a rare orthopedic condition. The deformity has different directions of the angulation: anterolateral, anteromedial, or posteromedial.[1] Each presentation type is commonly associated with different etiologies. Anteromedial bowing is associated with fibular hemimelia. Fibular hemimelia is the most common long-bone deficiency and its presentations cover a wide range of deformities, ranging from mild leg-length discrepancy to severe shortening of the affected limb with foot, ankle, and knee deformities.[2] Posteromedial bowing always presents with talipes calcaneus that resolves spontaneously within the first months. In most cases, spontaneous correction of the tibial deformity also occurs, to some extent, within the first four years of life, although leg-length discrepancies commonly require surgical management.[3]

Anterolateral tibial bowing is the presentation with the worst prognosis due to its risk of pathologic fracture and pseudarthrosis.[4] This type of bowing is commonly associated with CPT and NF1. The natural history of CPT is extremely unfavorable and once a fracture occurs, there is a little or no tendency for the lesion to heal spontaneously.[5]

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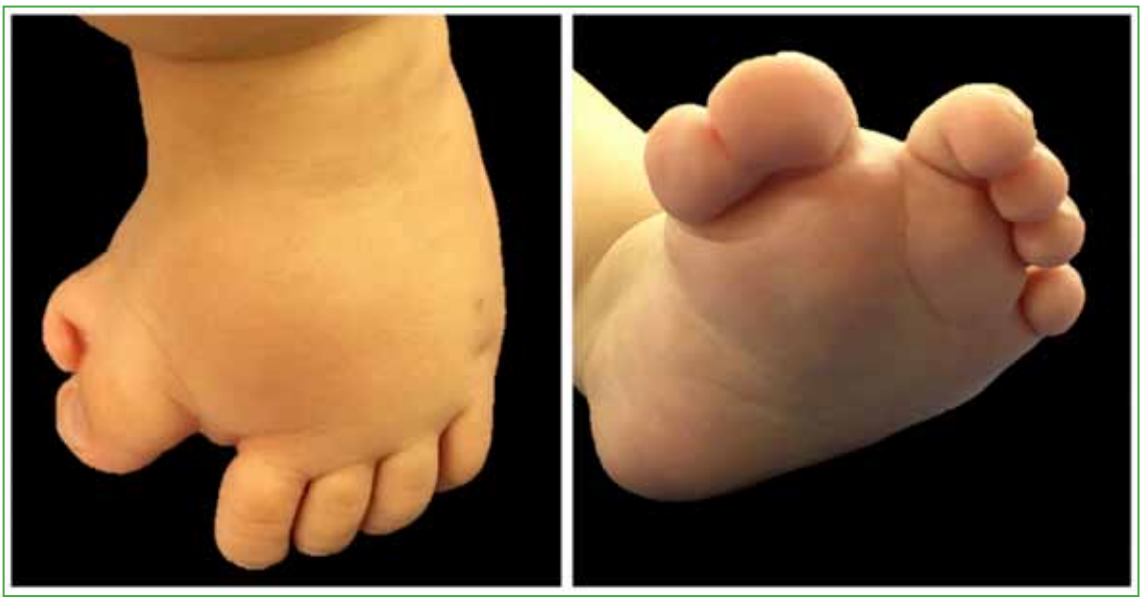
Although there is a strong relationship between anterolateral bowing of the tibia and CPT, there is an extremely rare presentation that has a more favorable prognosis, namely CABTP. Only 21 CABTP cases have been reported in the literature so far.[6-15]

We report a new CABTP case and our analysis of the literature CABTP cases in terms of associated deformities, diagnosis, and treatment.

CLINICAL CASE REPORT

A 6-month old boy was referred to our center Department of Pediatric Orthopedics for further evaluation and treatment of his left-foot polydactyly. The patient had no relevant family or personal history. Physical examination revealed preaxial polydactyly (left hallux) with complete phalangeal and metatarsal duplication. The medial supernumerary digit was hypoplastic and the interdigital space was widened (hallux varus) (Figure 1A).



A

Figure 1. Six-month-old boy. **A.** Photograph of the physical evaluation showing varus deformity of the hallux associated with preaxial polydactyly.

In addition, the patient presented with an anterolateral bowing of the tibia and a minor leg-length discrepancy. Physical examination revealed no café-au-lait spots in the skin nor any other signs of neurofibromatosis. Feet and lower limbs radiographs confirmed the presence of deformity (Figure 1B and C). The deformity association motivated a genetic study, which confirmed the CABTP diagnosis.

After discussing CABTP diagnosis and prognosis with the patient's parents, the decision was made to proceed with the surgical treatment of the foot deformity. The supernumerary toe was removed, and the realignment of the hallux varus was performed using the Farmer's technique to reduce the interdigital space. There were no complications during the surgery or in the postoperative period. Twenty-two months after surgery, the patient presented good evolution with adequate hallux alignment. Although the tibial deformity had a favorable course, the leg-length discrepancy increased (Figures 2 and 3). The patient currently wears a shoe lift and will require a surgical intervention to correct the leg-length discrepancy.

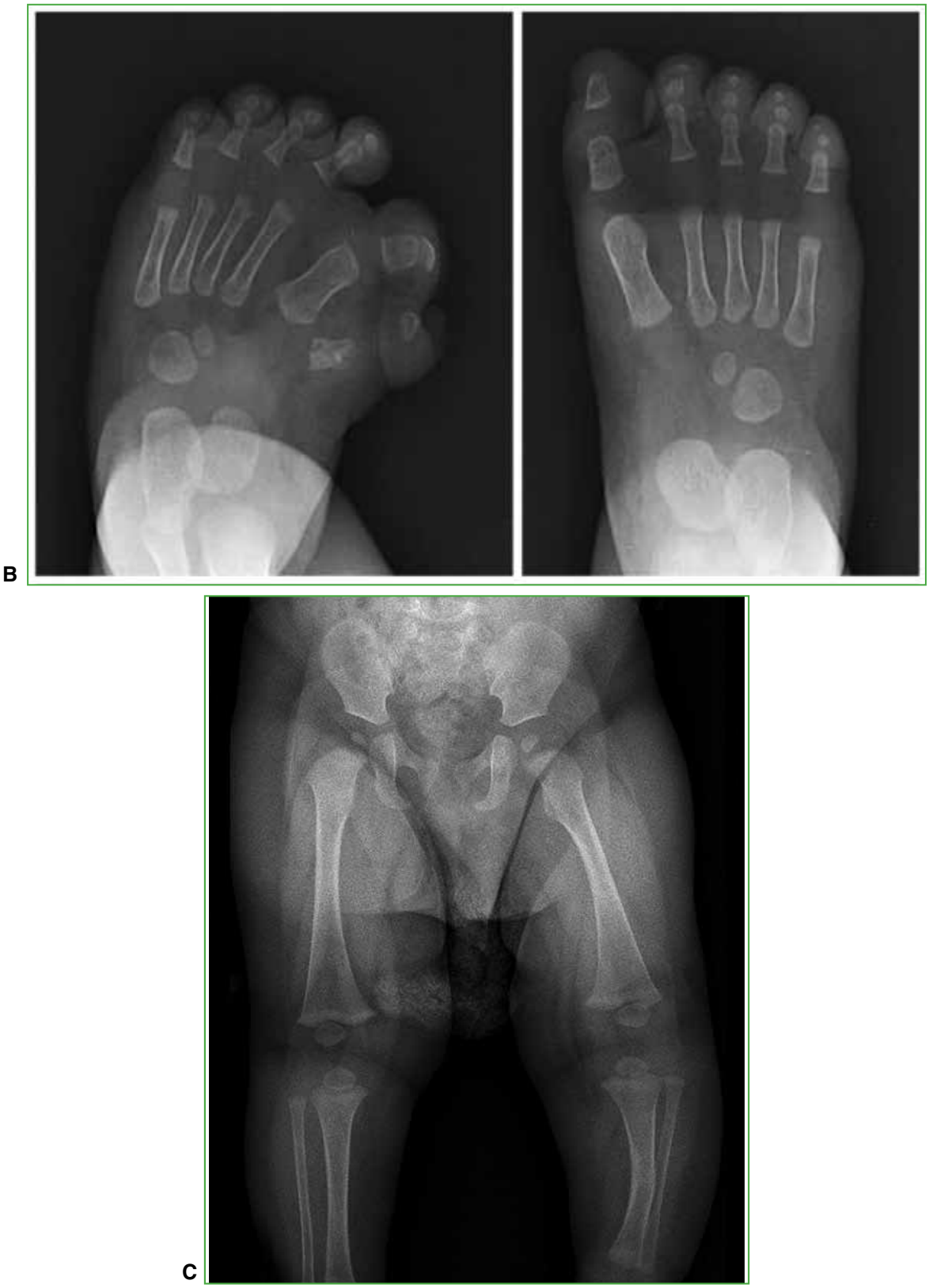


Figura 1. Six-month-old boy. **B.** Anterior-posterior radiographs of both feet confirming the preaxial polydactyly diagnosis. **C.** Lower limbs radiographs evidencing anterolateral bowing of the left tibia and mild leg-length discrepancy.

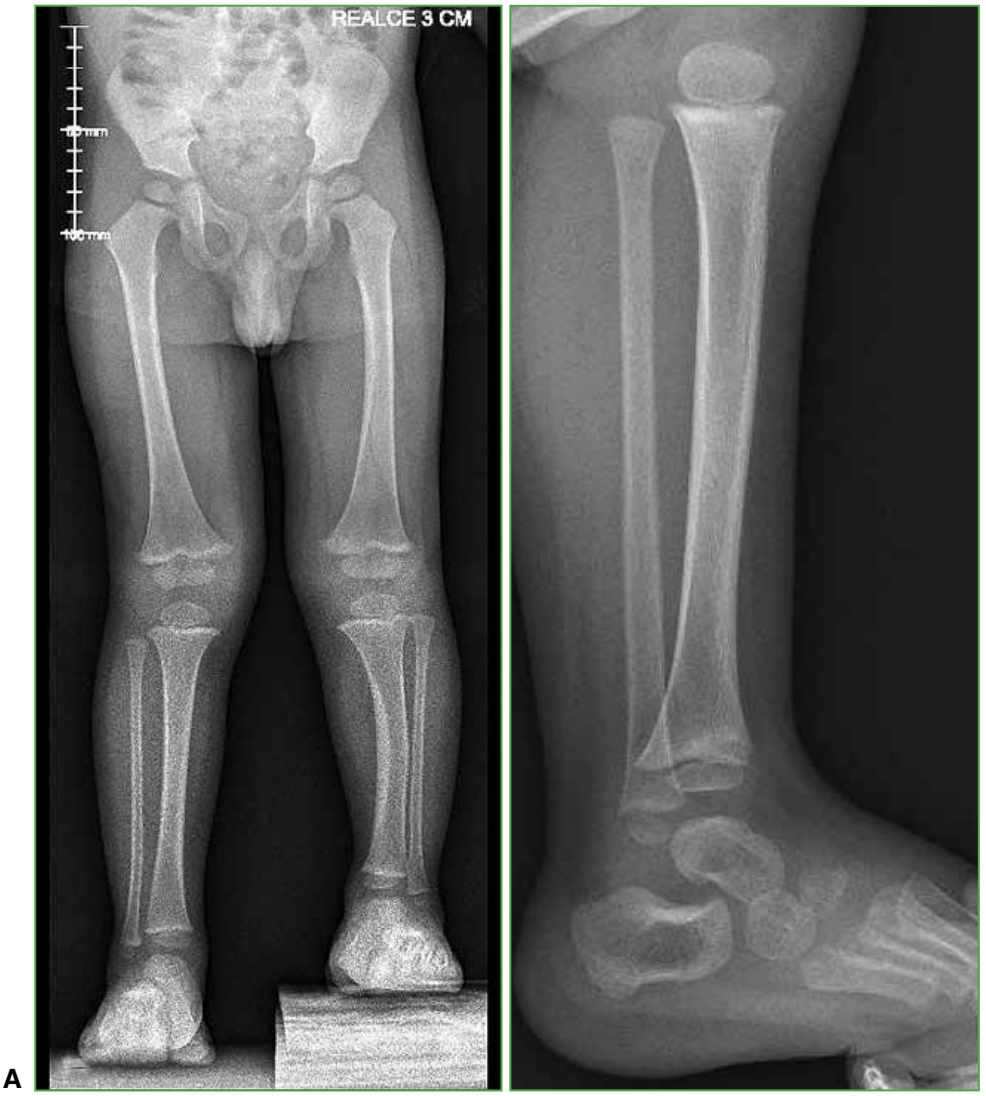
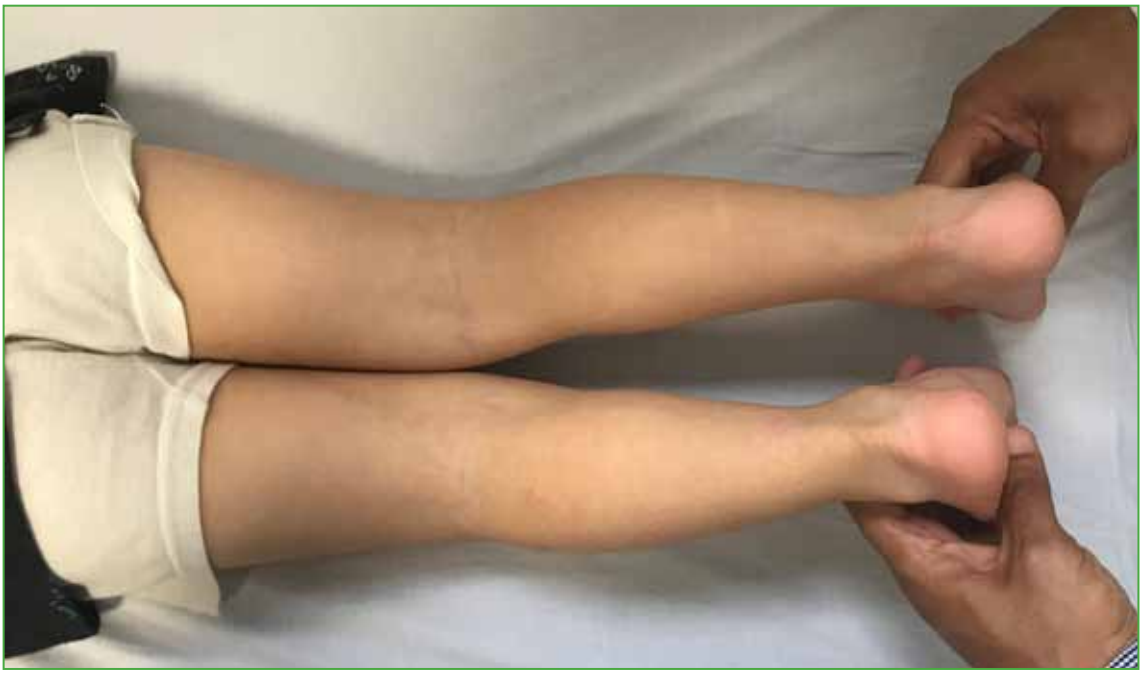


Figure 2. The same patient, 26 months later. **A.** Lower limbs radiographs evidencing a clear improvement regarding the anterolateral bowing of the left tibia, and an increased leg-length discrepancy (2.8cm). **A.** Left tibial lateral view radiograph showing no sign of deformity in the sagittal plane. **C.** Anteroposterior and lateral left foot radiographs taken 20 months after polydactyly correction.

Figure 3. Postoperative photographs showing the left foot smaller size (**A and B**).

C



D

Figure 3. Postoperative photographs showing the leg-length discrepancy (**C and D**).

DISCUSSION

The congenital anterolateral bowing of the tibia is a skeletal deformity that usually presents associated with NF1 and CPT.[1] CPT is a challenging condition because there is no chance of spontaneous correction, and, once pseudarthrosis has occurred, there is a very low chance for achieving bone union. Therefore, initial treatment is directed to prevent fracture and the progression of deformity by wearing orthosis.[16] Although their presentations are commonly associated, the anterolateral bowing of the tibia is not exclusively associated with CPT or NF1. There is a presentation associated with ipsilateral polydactyly of the hallux that has a more favorable prognosis. CABTP is a condition of unknown etiology which is diagnosed based on clinical and radiological findings. There are very few reported cases of CABTP (Table).

Table. Demographics of patients from the reported cases of congenital anterolateral tibial bowing associated with polydactyly

Author	Journal	n	Age*	Sex	Side	Associated deformities
Newell	*J Bone Joint Surg* (1976)	1	3 years	M	L	
Adamsbaum	*Pediatr Radiol* (1991)	5	17 months	F	L	Bilateral syndactyly, hallux duplication
			12 years	M	R	Bilateral clinodactyly
			10 years	M	L	Left clinodactyly
			7 months	M	R	Equinovarus foot, right clinodactyly, hallux hypertrophy
			6 years	M	R	
Weaver	*J Pediatr Orthop* (1996)	2	9 months	M	L	
			6 years	M	L	Duplication of the navicular, cuneiform, and metatarsal bones, radial deviation of both index fingers
Kitoh	*Am J Med Genet* (1997)	1	13 years	M	R	
			18 months		R	
Bressers	*J Pediatr Orthop B* (2001)	1	17 months	M	L	
Manner	*J Bone Joint Surg Br* (2005)	3	6 years	F	-	
			5 years	M	-	
			5 years	F	R	
Lemire	*J Med Case Rep* (2007)	1	2 years	M	R	Radial deviation of the left index finger
Breckpot	*Clin Dysmorphol* (2009)	2	19 years	M	R	Agenesis of the corpus callosum, interhemispheric cyst, mild syndactyly
			2 years	M	R	Agenesis of the corpus callosum, interhemispheric cyst, aqueduct stenosis, right clinodactyly, both index fingers distal hypoplasia
Han	*Am J Med Genet Am* (2012)	3	28 months	M	R	
			2 years	M	L	Mild fibula bowing
			2 years	M	L	
Beck	*J Pediatr Orthop B* (2013)	2	15 years	M	L	Equinovarus foot
			16 years	M	R	Equinovarus foot
This study		1	2 years	M	L	

F: female; L: left; M: Male; R: right.
*Age at last follow-up control.

In 1976, Newell and Durbin[6] described a type of anterolateral bowing of the tibia with no evidence of NF1, although with congenital deformations, that tended to correct spontaneously. In 1991, Adamsbaum *et al*.[7] established the association between the anterolateral bowing of the tibia and ipsilateral preaxial duplication of the hallux as a separate entity of favorable prognosis. More recently, Weaver *et al*.,[8] Bressers and Castelein,[10] and Manner *et al*.[11] published studies on similar cases, which included a more detailed description of the tibial morphological changes through radiographic and CT imaging studies.

CABTP patients commonly present with associated upper-limb deformities, including clinodactyly and polydactyly, as well as associated cerebral malformations, including agenesis of the corpus callosum. Although there is no distinctive association with hand and brain malformations, all patients had unilateral anterolateral bowing of the tibia and severe hallux varus with duplication of phalanges, first metatarsal bone or even tarsal bones, which was always ipsilateral to the tibial involvement.[6-14] A characteristic aspect of these patients is that the apex of the deformity is commonly located in the middle third of the tibia unlike pseudarthrosis patients, who commonly present the apex at a more distal level, at the junction of the middle and distal thirds of the tibia. CT findings at the apex of the tibial bowing vary in terms of morphological characteristics. Adamsbaum *et al*.[7] and Manner *et al*.[11] described the duplication of the tibial medullary canal, while Kitoh et al.[9] reported a posteromedial cortical cleft of the tibia at the site of the deformity. CABTP patients do not usually present with radiographic evidence of sclerosis, medullary canal narrowing or nonunion, which are characteristic features of pseudarthrosis. In addition, fibula involvement is also uncommon, although there may be a slight discrepancy or bowing in relation to the contralateral fibula. All the reported cases achieved spontaneous correction of the tibial bowing. Initial management is required to treat hallux duplication (often within the first months of life) and further treatment may be required to correct leg-length discrepancies.

Our patient had no hereditary history, associated deformities not fibula involvement. The radiographic follow-up showed adequate evolution with the tibial deformity correction, but an increasing leg-length discrepancy. As CT findings do not alter the treatment, the CT study was ruled out to avoid radiation exposure. Early surgical management of the preaxial polydactyly allowed for forefoot realignment and the use of conventional footwear without any difficulty. The leg-length discrepancy has been initially managed with the use of shoe lift, but surgery is required to equalize leg length.

CONCLUSION

CABTP is a condition of unknown etiology that includes unilateral anterolateral bowing of the tibia associated with ipsilateral duplication of the hallux. Diagnosis is based on clinical and radiological findings. The apex of the deformity is in the middle-third of the tibia (more proximal than in CPT patients) and there is a tendency for the deformity to correct spontaneously. This distinctive characteristic allows for avoiding unnecessary orthotic management and counseling the family on the more favorable prognosis of this condition.

Conflict of interests: Authors claim they do not have any conflict of interest.

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