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Symptomatic hallux valgus and dorsal bunion in adolescents with cerebral palsy: clinical and biomechanical factors

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ABBREVIATION

MTP Metatarsophalangeal

AIM The prevalence of severely symptomatic deformities of the first metatarsophalangeal (MTP) joint in adolescents with cerebral palsy (CP) requiring arthrodesis is unknown. Recent literature regarding these deformities is limited. We studied the presentation of severe, symptomatic deformities of the first ray in a large population of children and adolescents with CP and their association with gross motor function, CP subtype, and other musculoskeletal deformities.

METHOD We identified 41 patients with CP and a symptomatic deformity of the first MTP joint, managed by arthrodesis, from a large population based database over a 21-year period. Information recorded included demographics, CP subtype, Gross Motor Function Classification System (GMFCS), clinical presentation, and radiological features.

RESULTS Adolescents with spastic diplegia, at GMFCS levels II and III, were the most common group to develop symptomatic hallux valgus. In contrast, non-ambulant adolescents, at GMFCS levels IV and V, with dystonia or mixed tone, more commonly had dorsal bunions.

INTERPRETATION The type of first MTP joint deformity in patients with CP may be predicted by the type and distribution of movement disorder, and by GMFCS level. Specific patterns of associated musculoskeletal deformities may contribute to the development of these disorders and may provide a guide to surgical management.

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What this paper adds

- The prevalence of severe bunions requiring fusion surgery was 2 per cent.
- The two types of bunion were hallux valgus and dorsal bunion.
- The type of bunion can be identified on both clinical and radiological grounds.
- The cerebral palsy subtype is predictive of the type of bunion.

[main text]

Musculoskeletal deformities are common in cerebral palsy (CP) and may affect any part of the limbs and trunk.[1] Deformities vary with the type, severity, and distribution of the movement disorder, as well as the functional level of the patient. The risk of hip displacement and the shape of the proximal femur are closely related to the patient's Gross Motor Function Classification System (GMFCS) level.[2–4] Many other patterns of deformity have been recognized, including flexion contractures in both the upper and lower limbs, torsional deformities of long bones, and various malalignments of the foot and ankle.[1]

Deformities in the foot present as complex segmental malalignments which seem to result from the interplay between the movement disorder, muscle imbalance, contractures, and abnormal biomechanics.[5,6] Pes equinovarus is the most common foot deformity in patients with unilateral involvement, while pes valgus is more common in bilateral involvement.[5–8] Deformities of the first ray, including hallux valgus and hallux flexus (dorsal bunion), are not uncommon in adolescents with CP. Symptoms such as pain, brace intolerance, difficulties with footwear, gait disruption, and skin compromise may lead to the need for reconstructive surgery.[5,9–16] Despite the fact that these common deformities can cause debilitating symptoms, their pathogenesis and optimum management have not been fully elucidated. The description of normal radiographic

values in the pediatric foot has improved the understanding of these segmental malalignments.[17]

The purpose of this study was to study the prevalence and presentation of severe bunions, requiring surgery, in a large population of children and adolescents with CP. More specifically, our goal was to determine if differences in the movement disorder, topographical distribution, and functional levels affect the type of first metatarsophalangeal (MTP) deformity. A second goal was to determine which patterns of musculoskeletal deformity are associated with specific types of first MTP deformity.

METHOD

Ethical approval for this retrospective cohort study was given by the Ethics in Human Research Committee (institution 29151) and no external funding was received. A retrospective review of the orthopaedic department database, gait laboratory, and hip surveillance records and the state-wide Cerebral Palsy Register was carried out.[18] The criteria for inclusion were a diagnosis of CP, registration on the CP register, and symptomatic deformity of the first MTP joint, which was treated with arthrodesis of the first MTP joint between January 1995 and December 2016. Arthodesis is our procedure of first choice after failure of non-operative management including orthotic and footwear alterations.

Forty-one patients were identified who had surgery over the 21-year period, for a prevalence of 1.8 per cent of patients on the CP register. Clinical and radiological data were extracted from patient records and the patient's digital radiographs (Picture Archiving and Communication System).

The patients were classified by their functional level according to GMFCS, as well as their movement disorder (spastic, dystonic, and mixed tone or spastic-dystonia) and topographical distribution (hemiplegia, diplegia, triplegia, and quadriplegia).[18] In some patients, classification was noted to have changed with time. For example, at the time of registration, 15 to 20 years ago some children were classified as having 'spastic quadriplegia'. Later chart records confirmed the presence of dystonia or mixed tone. In such cases, the most up-to-date classification information was used.

After identification of the cohort, multiple sources were reviewed to extract additional clinical and radiological information including history of additional acquired

musculoskeletal problems. Ambulant patients who had undergone single event multilevel surgery were identified. Non-ambulant patients who had undergone surgery for hip displacement, spinal fusion, and any other orthopaedic procedures were identified.

All patients had standardized anteroposterior and lateral radiographs of the feet before first MTP fusion. Weight bearing views were obtained in all ambulant patients. In non-ambulant patients, simulated weight bearing views were obtained by having the patient seated in their wheelchair with the film cassette placed under the foot for the anteroposterior view or at the side for the lateral view. Measurements made on anteroposterior and lateral foot radiographs included hallux valgus angle, intermetatarsal angle, interphalangeal angle, and lateral MTP angle. These measures were performed on digital radiographs by previously described methods, which are reasonably familiar.[17] The first metatarsal declination angle is less well known and has rarely been described in paediatric foot disorders.[19] This is the angle between the long axis of the first metatarsal and the floor, on a lateral radiograph of the foot (Figure S1, online supporting information). The normal range in adult feet is approximately 20 degrees to 22 degrees,[20,21] but the normal range for the paediatric population has not been reported. A positive value denotes plantarflexion of the first metatarsal, with respect to the floor, and a negative value denotes dorsiflexion of the first metatarsal with respect to the floor. Radiological measurements were made on good quality digital radiographs on the Picture Archiving and Communication System by one attending orthopaedic surgeon and by one fellow in orthopaedic surgery. The mean of each pair of measurements was used for subsequent analysis.

The 41 patients were divided into two groups based on radiological features and in equivocal cases, the location of symptoms. These were predominantly on the medial side of the first MTP joint (hallux valgus) or on the dorsal aspect of the first MTP joint (dorsal bunion). The association between each of the two groups with other clinical and demographic factors was examined. The number of patients with each type of movement disorder, topographical distribution, and GMFCS level were determined within each group and compared using Fisher's exact test. Radiologic measurements for each group were analysed. Clinical and radiologic differences between the two patient groups were described using means, standard deviations (SD), and percentages. An unpaired Student's

t-test was used for the determination of statistically significant differences in the radiographic parameters between the hallux valgus and dorsal bunion group. *P*-values less than 0.05 were considered significant.

RESULTS

Patient demographics, gross motor function, movement disorder, and topographical distribution for the hallux valgus and dorsal bunion groups are summarized in Table I. The mean age at the time of first MTP arthrodesis was 15 years and 9 months (SD 1.6, range 12–18y) in the hallux valgus group, and 17 years (SD 1.8, range 15–20y) in the dorsal bunion group. The majority of patients with hallux valgus had spastic diplegia and were ambulant and functioning at GMFCS levels I to III. Patients with dorsal bunion were all non-ambulatory, functioning at GMFCS levels IV and V. Patients with unilateral hallux valgus had spastic hemiplegia and the majority of patients with bilateral bunions had spastic diplegia (or triplegia), were ambulant, and functioning at GMFCS levels I to III. Patients with dorsal bunions had dystonia or mixed tone, in a quadriplegic distribution, were non-ambulant, and at GMFCS levels IV and V.

The radiological data for each of the two groups is summarized in Table II. The hallux valgus angle was increased in both groups but was more severe in the hallux valgus group. The intermetatarsal angle and interphalangeal angle were increased in both groups. In radiographic terms, the biggest differences between the two groups were in the lateral MTP angle and the metatarsal declination angle. Not surprisingly, the hallux was more flexed in the dorsal bunion group compared to the hallux valgus group. In addition, the metatarsal declination angle was more dorsiflexed in patients with dorsal bunion than hallux valgus.

The majority of study patients had undergone surgery for correction of other musculoskeletal deformities. These procedures are presented in Table III. Ambulant patients had a high prevalence of procedures for equinus, pes valgus, lateral tibial torsion, and stiff knee gait. Non-ambulant patients had a high prevalence of procedures to prevent or treat hip displacement and scoliosis correction. No patients had prior surgery for their bunions, except one patient with a dorsal bunion. This patient developed skin breakdown,

deep infection, and required surgical debridement, antibiotics, and dressings to achieve healing and eradication of infection.

DISCUSSION

In our centre, the prevalence of CP is monitored using the state-wide Cerebral Palsy Register which has high levels of accuracy and case ascertainment.[2,18] In the past 20 years the prevalence of CP has remained relatively steady at approximately 120 new cases per annum. Throughout this period, approximately two patients per annum presented with symptomatic disorders of the first MTP joint which were considered to be severe enough to require a first MTP arthrodesis, for a rate of 1.8 per cent. In this study we had no information on patients with milder deformities that did not undergo first MTP arthrodesis, nor did we have information on the natural history of bunions later in adult life, in persons with CP.

Our review of children and adolescents with CP, who presented with symptomatic first MTP deformities, show a distinct separation into two main groups. Ambulant children, specifically those functioning in GMFCS levels II and III, with a predominantly spastic motor disorder, who previously had single event multilevel surgery to improve their gait and functioning tended to present as teenagers with symptomatic hallux valgus.[5,6] In contrast, non-ambulant children, in GMFCS levels IV and V, with either dystonia or spastic dystonia and a high prevalence of hip displacement and spinal deformity, presented with dorsal bunion. The separation into two groups according to where the symptoms were located was simple and unequivocal. Adolescents with hallux valgus complained of pain and callosities on the medial side of the first MTP joint. In contrast, teenagers with dorsal bunions presented with pain, inflammation, and skin breakdown on the dorsum of the first MTP joint. There was no overlap in clinical symptoms between the two groups. However, separation of the groups according to radiological features was less clear. For example, the majority of patients with a dorsal bunion on clinical grounds had some degree of hallux valgus. The most useful radiological parameter to separate the two groups was the metatarsal declination angle.

Hallux valgus is a common acquired deformity in patients with CP and may be multifactorial in origin. The association between hallux valgus and previous surgical

treatment for equinus, pes valgus, and cocontraction at the knee suggests that the previously suggested etiology, that is, that hallux valgus results from repeated 'toe scuffing' and laterally directed forces during the stance phase of gait, may be correct.[5,12–14] It is not known and cannot be determined from our study whether earlier treatment for some of these lower limb deformities, including equinus and pes valgus, might reduce the incidence of symptomatic hallux valgus.[5] We deferred surgical treatment for hallux valgus, even when it was present in younger children presenting for single event multilevel surgery, in an effort to see if the deformity and associated symptoms would resolve after successful correction of pes valgus and stiff knee gait. However, stabilization of the first MTP joint with arthrodesis, in combination with correction of coexisting malalignments in the lower extremity, appears to achieve good results for the majority of ambulant children.[5,16] In our centre, non-fusion surgery has resulted in poor outcomes and we prefer to delay until symptoms and deformity indicate a fusion procedure.

In contrast, less has been written about hallux flexus in CP. Although there have been many theories as to its pathogenesis, relatively little is known about this disorder. Until now, it has been believed to be the result of over-activity of the extensor muscles of the hallux and ankle, most often in patients having undergone previous orthopaedic procedures.[22] Iatrogenic dorsal bunions have been described as complications of various tendon lengthening and transfers.[13,23] Various surgical treatments have been advocated for dorsal bunion, including plantarflexion osteotomy of the first ray plus arthrodesis of the metatarsal-cuneiform and cuneiform-navicular joints, excision of the proximal portion of the proximal phalanx, various tendon transfers about the hallux, and arthrodesis of the first MTP joint.[13,23] Most research on dorsal bunions has been Level IV and V evidence, that is, small case series and case reports, on a heterogeneous patient population with various diagnoses. This is the first study to identify specific characteristics of patients with dorsal bunions in CP.

Our study shows a high prevalence of a dystonic movement disorder characterized by a withdrawal reflex and excessive flexion at the hip and knee as well as dorsiflexion at the ankle (Fig. 1). We hypothesize that the cascade of events leading to symptomatic dorsal bunion in adolescents with CP may be: (1) the emergence of dystonia or mixed

tone around the time of the adolescent growth spurt; (2) increased ankle dorsiflexion and decreased ankle plantarflexion secondary to dystonic over pull of the dorsiflexors compared to the plantar flexors; (3) progressive elevation of the first metatarsal; (4) progressive flexion at the first MTP joint. Ankle dorsiflexion and first metatarsal elevation exaggerate the tightness in both the short and long toe flexors; (5) pain, inflammation and skin breakdown over the dorsum of the prominent first MTP joint.

This cascade theory is hypothesis but fits the observed facts. It could only be confirmed by a longitudinal study of pathogenesis. In our patient population, the presence of a dorsal bunion was rarely recorded in clinical records, until it became symptomatic in the mid to late teens. Symptoms then progressed rapidly with a mean interval of 7 months between first presentation and fusion surgery.

Again, it cannot be determined from our data set and it is not known whether earlier and more vigorous treatment of dystonia in the tibialis anterior and calcaneus at the ankle might reduce the later prevalence of symptomatic hallux flexus.

There are limitations to our study. First, as previously noted, patients with first MTP deformity that did not undergo arthrodesis were not included in our study. As this is a retrospective review, we did not have means to identify these patients within our various databases and registries. Second, we did not employ a standardized scale to quantify preoperative symptoms. The modified American Orthopaedic Foot and Ankle Society Clinical Rating System Hallux Metatarsophalangeal-Interphalangeal Scale has been used in previous studies.[5,24,25] This scale has not been validated for patients with CP. The scale includes scoring for functional limitations and may therefore not be applicable to patients with major functional limitations, especially GMFCS levels IV and V. Thirdly, we described the features of adolescents presenting with severe, symptomatic deformities of the first ray of the foot in the second decade of life, who required a fusion procedure. We have no information of problems, which may occur later in life.

First MTP deformities are common in adolescents with CP. The development of specific deformities may be predicted by the distribution, type, and severity of movement disorder. The identification of these patient characteristics enhances our understanding of the pathogenesis, and may aid prevention and guide surgical management of these disorders.

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The authors have stated that they had no interests which might be perceived as posing a conflict or bias.

SUPPORTING INFORMATION

The following additional material may be found online:

Figure S1: Measurements on the lateral radiograph of the foot.

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[**FIGURE LEGEND]**

Figure 1: Illustration of the pathogenesis of a dorsal bunion (a) and clinical photographs of a patient with severe spastic dystonia, in Gross Motor Function Classification System level V; (b) showing the withdrawal reflex and excessive flexion at the hip and knee as well as dorsiflexion at the ankle, resulting in progressive elevation of the first metatarsal and flexion at the first metatarsophalangeal joint.

Table I: Patient characteristics

Variables	**Hallux valgus group** ***n*** **(%)**	**Dorsal bunion group** ***n*** **(%)**	**Fisher's exact test** ***p***
Gender			
Male	11 (52)	13 (65)	0.5303
Female	10 (48)	7 (35)	
GMFCS			
I	2 (10)	–	<0.0001[a]
II	10 (48)	–	
III	7 (33)	–	
IV	2 (10)	8 (40)	

V	–	12 (60)	
Movement disorder			
Spastic	18 (86)	–	<0.0001[a]
Spastic-dystonia	3 (14)	12 (60)	
Dystonia	–	8 (40)	
Topographical distribution			
Hemiplegia	3 (14)	–	<0.0001[a]
Diplegia	15 (71)	–	
Triplegia	1 (5)	2 (10)	
Quadriplegia	2 (10)	18 (90)	

[a]*p*<0.0001 between proportions GMFCS levels I–III and GMFCS levels IV–V, spastic and spastic-dystonia/dystonia, and hemiplegia–diplegia–triplegia and quadriplegia of hallux valgus group and dorsal bunion group, using Fisher's exact test. GMFCS, gross motor function classification system.

Table II: Radiographic measurements for the right and left feet in the hallux valgus and dorsal bunion groups

Variables	**Hallux valgus group**	**Dorsal bunion group**	**Unpaired *t*-test**
	Degrees	**Degrees**	***p***
HVA			
Right	41 (12)	31 (16)	0.0262*

Left	41 (10)	32 (15)	0.0189*
IMA			
Right	12 (5)	12 (3)	0.5805
Left	12 (4)	12 (3)	0.6664
IPA			
Right	13 (7)	10 (6)	0.2382
Left	13 (7)	11 (4)	0.2393
L-MPTA			
Right	5 (17)	52 (10)	<0.0001[a]
Left	4 (17)	52 (9)	<0.0001[a]
MDA			
Right	19 (5)	-1 (6)	<0.0001[a]
Left	19 (5)	-1 (6)	<0.0001[a]

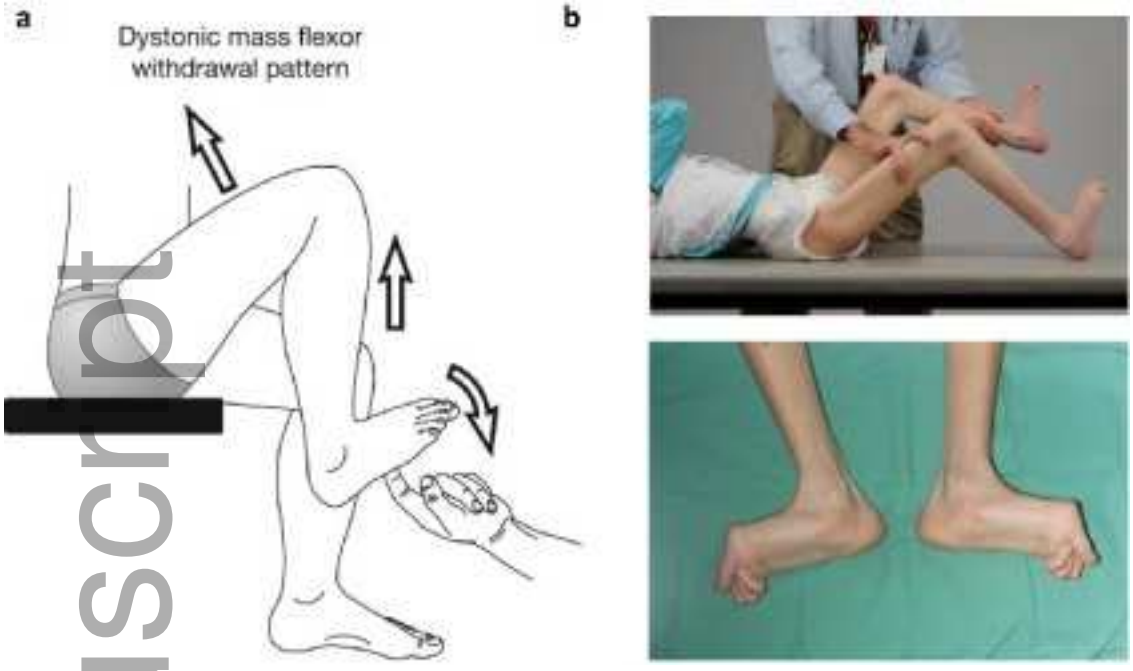
The values are given as the mean (± standard deviation). [a]*P*-values less than 0.05 were considered significant. HVA, hallux valgus angle; IMA, intermetatarsal angle; IPA, interphalangeal angle; L-MTA lateral metatarsophalangeal angle; MDA, first metatarsal declination angle.

Table III: Overview of procedures for correction of other musculoskeletal deformities preceding the arthrodesis of the first metatarsophalangeal joint

Procedures	***n***
Hallux valgus group	

Surgery for equinus contracture	21
BoNT-A for spastic equinus	18
Surgery for pes valgus	17
Rectus femoris transfer	13
Supramalleolar osteotomy	12
Femoral derotation osteotomy	10
Total	**91**
Dorsal bunion group	
Hip surgery[a]	18
BoNT-A	14
Scoliosis surgery	11
Baclofen pump	5
Total	**48**

[a]Hip surgery included adductor releases in 18 patients with bony reconstructions in 15 patients. BoNT-A, botulinum neurotoxin A.



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