Childs Nerv Syst (2016) 32:315–319
DOI 10.1007/s00381-015-2944-7

ORIGINAL PAPER

Incidence and type of foot deformities in patients with spina bifida according to level of lesion

Huseyin Gunay[1] · **Murat Celal Sozbilen**[1] · **Yusuf Gurbuz**[2] · **Mahmut Altinisik**[1] · **Beyhan Buyukata**[3]

Received: 26 September 2015 / Accepted: 22 October 2015 / Published online: 30 October 2015
© Springer-Verlag Berlin Heidelberg 2015

Abstract
Aim The previously suggested association between the incidence of high-level foot deformity and muscle imbalance is no longer supported, when evaluated independent from motor and sensory loss and level of lesion, by current studies. The purpose of this study was to evaluate the association between level of lesion and foot deformity.
Methods Of 545 patients, a total of 136 (272 feet) patients admitted to the spina bifida clinic between 2010 and 2014 were included in this study. Levels of all lesions were evaluated using initial operation data, the motor-sensory exams, and direct radiography. All patients were categorized into four different groups: Thoracic region (group 1), high-level lumbar—L1-2 region (group 2), mild and lower lumbar regions (L3-4-5) (group 3), and Sacral region (group 4).
Results The mean follow-up time was 34.9 months (range 8–176 months). Group 1, group 2, group 3, and group 4 included 24 (17.6 %), 14 (10.3 %), 19 (14 %), and 79 (58.1 %) patients with regards to level of lesion, respectively. The incidences of foot deformity were 85.4, 85.7, 81.5, and 50.6 % in groups 1, 2, 3, and 4, respectively. Of all patients, 22 % (61 feet) had clubfoot, 16 % (44 feet) pes cavus, 10 % (26 feet) pes valgus, 6 % (17 feet) isolated equinus, 6 % (17 feet) pes calcaneus, and 5 % (13 feet) metatarsus adductus. Patients without a foot deformity (81 % of normal feet) usually had a lesion at the sacral level ($p \leq 0.05$). On the other hand, isolated equinus (70 %) and clubfoot (49 %) deformities were mostly observed in spinal lesions ($p > 0.05$). The incidence of pes calcaneus, pes valgus, and adductus deformities inclined as the lesion level decreased ($p > 0.05$).
Conclusion In this study, it was concluded that foot deformities were directly related to the level of lesion. The comparison of higher and lower level lesions revealed that the types of foot deformity differed significantly. The muscle imbalance due to spina bifida was not sufficient to explain the pathology. On the other hand, the level of spinal lesion is an important factor for the type of deformity.

Keywords Meningomyelocele · Foot deformities · Spina bifida occulta · Spina bifida cystica

Introduction

The main goal of treatment for spina bifida is to allow patients to achieve walking ability or maintain their current level of functioning. Besides hip and knee stability, the foot should be stable during the stance phase of gait and the swing phase should be consistent [1]. The high incidence of foot deformity in patients with spina bifida was, in the beginning, attributed to muscle imbalance, which was later contradicted by the results from studies reporting the presence of foot deformity despite no motor activity in patients with high-level lesions [2, 3]. On the other hand, it has been demonstrated that the incidence and types of foot deformity also vary depending on level of lesion and are independent of motor and sensory loss [4].

The purpose of this study was to evaluate the association between foot deformity and level of lesion in patients with spina bifida.

✉ Murat Celal Sozbilen
muratcelal@hotmail.com

[1] Department of Orthopaedics and Traumatology, Ege University, Izmir, Turkey

[2] Hand Microsurgery Orthopaedic Traumatology Hospital (EMOT), Izmir, Turkey

[3] Mus, Bulanık State Hospital, Mus, Turkey

Materials and methods

Of 545 patients who were admitted to the spina bifida clinic and had medical records in a meningomyelocele study group consisting of neurosurgery, pediatric urology, physical therapy rehabilitation, and orthopedics; a total of 136 patients (272 feet) with complete data and follow-up were enrolled in this study. The exclusion criteria were patients who were not operated on in the early period and those who had an unknown level of lesion. Patients' level of lesion and physical examination results were evaluated by two different experienced physicians; an orthopedist and a senior orthopedic resident. Lesion levels were assigned according to initial operation results, motor-sensory examinations, and direct radiographs. Patients were divided into four groups according to their lesion levels: thoracic region (group 1), high-lumbar region L1–2 (group-2), mid-lower lumbar region L3-4-5 (group 3) and sacral region (group 4). Data were obtained from medical records of the meningomyelocele study group which had been recorded once in every 6 months for 2 years after birth and subsequently annually and analyzed using SPSS 18.0 (SPSS Inc., Chicago, IL, USA).

Results

The mean age of the patients was 8.63 years (range 2 years, 1 month–22 years, 2 months); 69 (50.7 %) patients were males and 67 (49.3 %) were females. The mean follow-up time was 34.9 months (range 8–17 months). According to the lesion levels, group 1 (thoracic) included 24 (17.6 %) patients, group 2 (upper lumbar) 14 patients (10.3 %), group 3 (mid-lower lumbar) 19 patients (14 %), and group 4 (sacral) 79 patients (58.1 %). A total of six types of deformities were found and the incidence of deformity was 63 % (87 feet) for the right foot and 65 % (89 feet) for the left foot. The deformities were symmetric in 136 (77 %) of 176 feet. One or more operations were performed in 94 (53 %) feet due to deformities and recurrence throughout the follow-up period. The evaluation of the incidence of deformity according to the lesion level revealed that 85.4 % had lesions at the thoracic level, 85.7 % at the upper lumbar level, 81.5 % at the mid-lower lumbar level, and 50.6 % at the sacral level. Of all cases, 96 (35 %) had normal feet whereas 22 % (61 feet) had clubfoot, 16 % (44 feet) had cavus, 10 % (26 feet) had pes valgus, 6 % (17 feet) had isolated equinus and pes calcaneus, and 5 % (13 feet) had metatarsus adductus deformities (Table 1). Normal foot was more common in sacral level lesions (81 % of normal feet) ($p<0.05$). Pes cavus deformities were mostly found in low-level lesions, particularly at the sacral level ($p<0.05$). Isolated equinus (70 %) and clubfoot deformities (49 %) were more common in high-level spinal lesions ($p<0.05$). Pes calcaneus, pes valgus, and adductus deformities were less common in high-level lesions, whereas as the level of spinal lesion decreased, the incidence of these deformities increased ($p<0.05$).

Discussion

The incidence of spina bifida has been reported to be 2/10,000, though it differs depending on the developmental status of a country [5]. Previous studies in our region reported the incidence of spina bifida to be 1.9/1000, which however was later suggested to have reached up to 8.9/1000 after the Chernobyl nuclear disaster [6]. Forty percent of the lesions in spina bifida are found at the lumbosacral-sacral level and 26 % at the thoracolumbar level [7]. In this study, most of the patients had lesions at the sacral and lumbosacral levels.

The incidence of congenital foot deformities in neuromuscular diseases is high [8]. The pathology of foot deformities in spina bifida is yet to be clarified. Even though muscle imbalance and/or reflex spasticity in the intrauterine period and intrauterine positioning have been held responsible for the etiology of spina bifida, 75 % of deformities occur in the absence of spasticity. No evidence has been provided to confirm the hypothesis that foot deformities in patients with spina bifida are caused by intrauterine muscle imbalance or positioning [4, 9]. In a study of fetal cadavers with spina bifida, Ömeroğlu et al. reported that muscle fiber size and fibrosis in clubfoot deformities were significantly lower than those of the control groups in all foot muscles, except the gastrocnemius muscle. It also was demonstrated in their study that fibrosis was most evident in the peroneus longus muscle in those with high-level spina bifida. The muscle imbalance between antagonist muscle groups due to significant muscular denervation atrophy caused talipes equinovarus (TEV) deformity in those with high-level myelomeningocele. Relative excessive activity of the plantar flexor and invertor muscles can lead to TEV deformity in patients with meningomyelocele [10].

It is well known that 75 % of patients with spina bifida have foot deformities [9]. Foot deformities vary depending on the lesion level, and as the lesion level decreases, the incidence of normal foot increase. Even though most of the cases presented in this study had spinal low-level lesions, 65 % had foot deformities. A literature review of previous studies of foot deformities in patients with spina bifida revealed four common types of foot deformities and that isolated equinus deformity was discussed with clubfoot deformities, whereas cavus deformities were handled with calcaneus deformity in most studies [2, 4, 11].

A study of patients with low-level spina bifida by Frawley et al. reported foot deformities in 263 of the 348 feet. Accordingly, of all feet, excluding normal feet, 33 % had equinus, 41 % calcaneus, 16 % valgus, 7 % varus, and 3 % convex pes valgus deformities [4]. In this study, 176 foot deformities were

Table 1 The types and number of foot deformities according to level of lesions in patients

Deformity level	N		Equinus		CF		PC		PPV		Cavus		Adduct		Total
	R	L	R	L	R	L	R	L	R	L	R	L	R	L	
Group 1	4	3	5	5	9	10	0	0	2	2	4	2	1	1	48
Group 2	2	2	2	0	5	6	1	1	2	2	2	1	0	2	28
Group 3	3	4	0	0	7	8	3	4	1	0	5	3	0	0	38
Group 4	40	38	3	2	7	9	3	5	8	9	14	13	3	4	158
Total	49	47	10	7	28	33	7	10	13	13	25	19	4	7	272
	96		17		61		17			26	44			11	

Group 1=Thoracic, Group 2=Upper extremity, Group 3=Mid-Lower Lumbar, Group 4=Sacral

N normal, *CF* clubfoot, *PC* pes calcaneus, *PPV* pes plano valgus, *Adduct* metatarsus adductus

found in 272 feet; 34.7 % clubfoot, 25 % cavus, 14.8 % valgus, 9.6 % isolated equinus, 9.6 % calcaneus, and 6.3 % adductus.

In this study, the incidence of pes calcaneus deformity was lower than that reported in the literature; however, when analyzed taking feet with cavus deformity into consideration, the incidence of pes calcaneus deformity was 34.6 %, which is also consistent with the literature data. The high incidence of pes calcaneus deformity was attributed to the fact that patients with sacral spina bifida constituted half of all patients. The high incidence of calcaneus and cavus deformities at lower levels is associated with the insufficiency due to innervation of the muscle groups (soleus, gastrocnemius), which are the antagonists of well-functioning tibial muscles innervated from the lumbar level, from the sacral region. In this study, the abundance of these deformities in the sacral region was statistically significant ($p \leq 0.05$). The incidence of all deformities, except that of the calcaneal deformity in the sacral region, did not differ among the levels [4]. In the treatment of pes calcaneus deformity, soft tissue surgical procedures usually fail to yield satisfactory results, and the release of the dorsal muscles of the foot, as well as the transfer of the tibialis anterior muscle to the calcaneus, arthrodesis, or calcaneal osteotomies constitute surgical treatment options. The ignorance of calcaneal deformity mostly results in heel ulcers and osteomyelitis (Fig. 1).

In this study, clubfoot and equinus deformities constituted 44 % of all deformities. The high incidence of equinus deformity was also reported in a study by Frawley et al., which included only patients with low-level spina bifida, unlike our study, in which, all levels of spina bifida were analyzed [4].

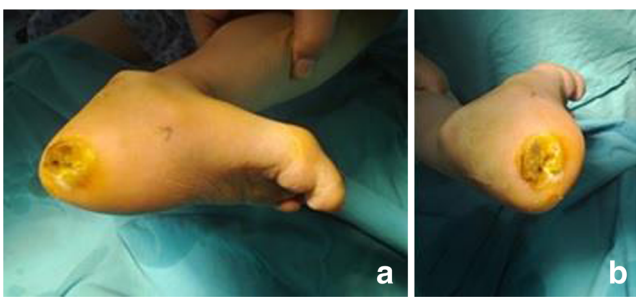

Fig. 1 **a**–**b** Foot ulcer in a patient with pes calcaneus deformity

Many factors including spasticity, intrauterine positioning, contractures, and muscle imbalance can contribute to the development of clubfoot in patients with spina bifida. For instance, contracture of the tibialis muscles or retained activity in combination with the function loss of the peroneal muscles may result in the development of clubfoot in low-lumbar level lesions [12].

The high incidence of equinus and clubfoot deformities in high-level spina bifida has been reported to be associated with flaccid paralysis of lower extremities [2]. On the other hand, there are large series in the literature reporting that equinus and club foot deformities are not associated with the lesion level [4]. In this study, even though clubfoot and equinus were more common in those with high-level lesions, no significant difference was found in their relationship with the lesion level ($p > 0.05$). In a study of 182 feet with all levels of spina bifida by Frischhut et al., 36 % of the feet had a clubfoot, 26 % had normal feet, 19 % had an equinus, and 17 % had a calcaneal deformity [11]. It was also reported that clubfoot was diagnosed in 90 % of patients with paraplegia due to a lesion at the high-level spinal area and 50 % in patients with paraplegia caused by a lesion at the sacral level. Clubfoot usually constitutes a risk for the development of pressure sores on the dorsolateral aspect of the foot. In addition, repeat surgery due to muscle imbalance was required in 50 % of these cases after primary correctional surgery [13]. Ankle-foot orthosis (AFO) following the lengthening of the Achilles tendon in patients with isolated equinus deformity and passive stretching exercises usually yield successful results; however, despite surgical release and tendon transfer in clubfoot deformity, which is usually more rigid in patients with spina bifida, the continuing growth of the child is associated with the increased incidence of recurrence, as a result of which, radical procedures such as talectomy as well as osteotomies including arthrodesis, lateral column shortening, or medial lengthening may be required (Fig. 2).

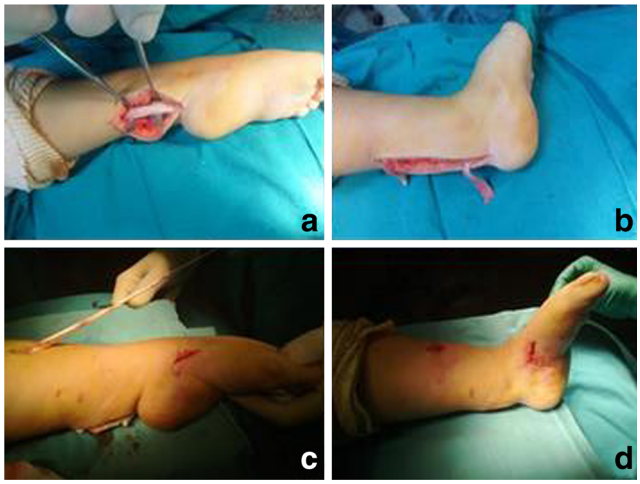

Fig. 2 **a**–**b** Correction of equinus by Achilles tendon lengthening. **c**–**d** Tibialis anterior tendon transfer in the same patient

Foot abnormalities were noted in 4 % of the infants, with 1 % having metatarsus adductus [14]. Widhe reported adductus deformity as the most common foot deformity at 3.1 % in infants [15]. In this study, even though the incidence of adductus deformity in patients with spina bifida did not differ greatly from that in normal population and was not associated with the lesion level, four patients had rigid deformities.

In this study, 15 % of the patients had a valgus deformity. It is most common in patients with L4 and L5 level lesions caused by strength or spasticity of the foot dorsiflexors combined with weak plantaris muscle. The predominance of the ankle evertors and lateral extensors of the foot over the anterior and posterior tibialis leads to a calcaneovalgus deformity [13, 16].

This deformity was more common at the lombosacral level in our series but was not associated with the lesion level ($p>0.05$). Even though patients with calcaneal deformity are recommended to undergo transfer of the tibialis anterior muscle to the calcaneus in the early period, secondary surgery may be required due to frequently encountered equinus deformity in the future. It has been reported that, with advancing age, anterolateral release of the ankle and opening wedge osteotomy of the calcaneus in combination with peroneal tendon lengthening yielded satisfactory results [17, 18]. In the study presented here, patients with valgus deformities usually underwent soft tissue release in combination with Grice-Green extraarticular subtalar arthrodesis and the Evans calcaneal osteotomy, and, of all foot deformities associated with spina bifida, more satisfactory results in terms of recurrence were obtained in patients with valgus deformities. In vertical talus deformity, which commonly leads to valgus deformity, talonavicular reduction is achieved using the K-wire joystick technique in the early period.

In this study, most of the patients with complete follow-up data had sacral and lumbosacral lesions. All spina bifida patients with sacral and lumbosacral lesions attended their first follow-up visit, whereas the follow-up attendance rate decreased throughout the study period, resulting in lower rates in this study. The nonattendance at follow-up appointments was mostly caused by the fact that patients with high-level lesions ended up wheelchair-bound because of the difficulty using HKAFO and reciprocating orthosis and that the treatment-related expectations of patients and their families declined in time. The high recurrence rate of deformities despite surgical procedures performed in these patients deepened the despair of patients and their families. On the other hand, patients with low-level lesions holding higher expectations for an active social life resulted in their strict attendance at follow-up visits.

Conclusion

The current approach suggests that muscle imbalance is not sufficient to explain the type of deformity in patients with spina bifida. The recurrence of deformities associated with advancing age, despite early treatment, indicates that the disease does not originate only in the intrauterine period. The aim of this study was to demonstrate the direct relationship between foot deformity and the lesion level in these patients. The comparison of high-level and low-level lesions revealed differences between the types of deformity. Accordingly, muscle imbalance involved at various levels due to spina bifida is not alone sufficient to explain the pathology; however, we believe that the spinal lesion level is an important factor for the type of deformity.

Compliance with ethical standards

Conflict of interest Each author certifies that he or she, or a member of his or her immediate family, has no funding or commercial associations (e.g., consultancies, stock ownership, equity interest, patent/licensing arrangements, etc.) that might pose a conflict of interest in connection with the submitted article.

References

1. Whittle, Michael W. Gait analysis: an introduction. 2003.
2. Broughton Nigel S, Geoffrey G, Menelaus MB (1994) The high incidence of foot deformity in patients with high-level spina bifida. J Bone & Joint Surgery, British 76(4):548–550
3. Dogan A, Albayrak M, Akman YE, Zorer G (2004) The results of calcaneal lengthening osteotomy for the treatment of flexible pes planovalgus and evaluation of alignment of the foot. Acta Orthop Traumatol Turc 40(5):356–366

4. Frawley PA, Broughton NS, Menelaus MB (1998) Incidence and type of hindfoot deformities in patients with low-level spina bifida. J Pediatr Orthop 18(3):312–313
5. Boulet SL, Gambrell D, Shin M, Honein MA, Mathews TJ (2009) Racial/ethnic differences in the birth prevalence of spina bifida—United States, 1995–2005. J Morb Mortal Wkly Rep 57(53):1409–1413
6. Çaglayan S, Kayhan B, Mentesoglu S,Aksit S;Changing incidence of neural tube defects in Aegean Turkey Paediatric and Perinatal Epidemiology Volume 3, Issue 1, pages 62–65, January 1989
7. BN French Midline fusion defects and defects of formation JR Youmans (Ed.), Neurological Surgery (2nd ed), W.B. Saunders, Philadelphia (1982), pp. 236–380 vol 3
8. Delpont, M., Lafosse, T., Bachy, M., Mary, P., Alves, A., & Vialle, R. (2014). [Congenital foot abnormalities.]. Archives de pediatrie: organe officiel de la Societe francaise de pediatrie.
9. Pediatric Orthopaedics: Core Knowledge In Orthopaedics; John P. Dormans, MD Philedelphia 2005 pp 492–493
10. Ömeroglu S, Peker T, Ömeroglu H, Gülekon N, Mungan T, Danisman N (2004) Intrauterine structure of foot muscles in talipes equinovarus due to high-level myelomeningocele: a light microscopic study in fetal cadavers. J Pediatr Orthop B 13(4):263–267
11. Frischhut B, Stöckl B, Landauer F, Krismer M, Menardi GJ (2000) Foot deformities in adolescents and young adults with spina bifida. Pediatr Orthop B 9(3):161–169
12. Flynn JM, Herrera-Soto JA, Ramirez NF, Fernandez-Feliberti R, Vilella F, Guzman J (2004) Clubfoot release in myelodysplasia. J Pediatr Orthop B 13:259–262. doi:10.1097/01.bpb.0000124491.13918.b7
13. Michael A, Bjoern B, Seyler TM, Wolfram W, Thomas B, Rainer A, Claus C (2009) Management of orthopaedic sequelae of congenital spinal disorders. J Bone Joint Surg Am 91(Supplement 6):87–100
14. Widhe T, Aaro SE, Elmstedt E (1988) Foot deformities in the newborn—incidence and prognosis. Acta Orthopaedica Scand 59:176–179
15. Widhe T, Foot Deformities at Birth (1997) A longitudinal prospective study over a 16-year period. J Pediatric Orthop Issue 17(1):20–24
16. Rodrigues RC, Dias LS (1992) Calcaneus deformity in spina bifida: results of anterolateral release. J Pediatr Orthop 12(4):461–464. doi:10.1097/01241398-199207000-00008
17. Park KB, Park HW, Joo SY, Kim HW (2008) Surgical treatment of calcaneal deformity in a select group of patients with myelomeningocele. J Bone Joint Surg Am 90(10):2149–2159
18. Erşahin Y (2013) Split cord malformation types I and II: a personal series of 131 patients. Childs Nerv Syst 29(9):1515–1526