Original Clinical Article

JOURNAL OF CHILDREN'S ORTHOPAEDICS

Additional challenges in children with idiopathic clubfoot: is it just the foot?

E. Lööf[1,2]

Abstract

Purpose Treatment of idiopathic clubfoot (IC) has improved since the introduction of the Ponseti method. However, relapses are still common and primarily related to non-adherence to the brace regime. Our hypothesis was that IC might be more than just a structural deformity. Based on three studies, the aim of this paper was to provide an overview of findings regarding additional challenges within IC.

Methods In total, 153 children with IC and 137 control children participated in the studies. The first study assessed gross motor skills in six motor tasks using the Clubfoot Assessment Protocol. The second and third studies surveyed neurodevelopmental difficulties (NDDs) using the Five to Fifteen (FTF) questionnaire and health-related quality of life (HRQoL) using the EuroQol-5D youth.

Results A high percentage of gross motor deviations were found in children with IC compared with controls, and those correlated poorly with clubfoot severity and foot movement. Children with IC had a higher prevalence of NDDs on the FTF compared with the control group, including the domains: motor skills, perception and language. One-third of children with IC were defined as at risk of developmental disorders. In this subgroup, parents were less satisfied with the outcome of clubfoot treatment and the children reported worse HRQoL than those without NDDs.

Conclusion The findings suggest additional challenges in children with IC, such as NDDs, apparently affecting both clubfoot treatment outcome and HRQoL. Thus, awareness of these challenges could be vital to further optimize treatment and support, for example, with regards to brace adherence.

Level of evidence II - Prognostic study

[1] Paediatric Neurology, Department of Women's and Children's Health, Karolinska Institutet, Karolinska University Hospital, Stockholm, Sweden
[2] Functional Area Occupational Therapy & Physiotherapy, Allied Health Professionals Function, Karolinska University Hospital, Stockholm, Sweden

Correspondence should be sent to Elin Lööf, Paediatric Neurology QB:27, Department of Women's and Children's Health, Karolinska Institutet, 171 76 Stockholm, Sweden.
E-mail: elin.loof@ki.se

Cite this article: Lööf E. Additional challenges in children with idiopathic clubfoot: is it just the foot? *J Child Orthop* 2019;13: 245-251. DOI: 10.1302/1863-2548.13.190076

Keywords: clubfoot; neurodevelopmental difficulties; health-related quality of life; gross motor skills; treatment outcome

Introduction

The treatment of idiopathic clubfoot (IC) has progressed since the global recognition of the Ponseti treatment method, now considered the benchmark treatment.[1,2] Examples of successful factors include a high initial correction rate and improved foot morphology, gait and patient-reported outcomes compared with surgical management.[2-4] Despite this, relapses of the deformity are still common and have been reported in up to 68% of cases.[5] Non-adherence to the brace regime is the constant and most prominent risk factor reported.[6] Because of this, many attempts have been made to increase brace compliance. Such attempts have included educational interventions, different brace designs and protocols.[7,8] Nevertheless, adherence to the brace regime can still be a challenge for families and an objectively measured daily brace use has revealed a median use of only 62% of the recommended time.[9]

Now that the Ponseti treatment method is considered the first-hand option, it is time to take the next step in the research and understanding of clubfoot. Although the pathogenesis of clubfoot is still unknown, progress has been made in understanding some of the genetic and environmental factors related to the deformity.[10-13] By identifying factors related to clubfoot as well as generating knowledge of the group of children with clubfoot, treatment and care can advance even further.

At Karolinska Institutet and Lund University, Sweden, our research team has been working on expanding the knowledge of children born with IC beyond the musculoskeletal deformity of the foot and lower leg itself. This work was based on the hypothesis that IC is more than just a structural problem. From the literature, we noted a finding made by chance where half the sample of boys with clubfoot, acting as reference material, were identified with attention-deficit/hyperactivity disorder (ADHD).[14] Moreover, another study found delays in attaining gross

motor milestones in infants with IC compared with typically developing infants, primarily with regards to independent walking.[15] Delays were not related to the treatment method used or initial severity of the clubfoot deformity. Interestingly, the authors suggested that one explanation could be that the clubfoot disorder might be a marker for an underlying mild motor development dysfunction. Similarly, Andriesse et al[16] found that low motor ability in children with IC at age seven years was not directly related to the status of the clubfoot, indicating additional difficulties besides the structural abnormality. Finally, in a study of children with idiopathic toe walking at an orthopaedic clinic, a higher prevalence of neurodevelopmental difficulties (NDDs) were reported,[17] implying a possible connection between NDDs and distal musculoskeletal deficits. In addition, our own clinical experiences have indicated underlying problems such as hyperactivity, language and attention in children with IC. Therefore, the combination of research findings aligned with indications from clinical practice led us to the hypothesis that additional challenges may occur within the group of children born with IC. The aim of the current paper was to present an overview of the findings based on the following studies conducted by our research team:

– Study 1: Lööf et al (2017),[18] Gross Motor Skills in Children with Idiopathic Clubfoot and the Association between Gross Motor Skills, Foot Involvement, Gait and Foot Movement.
– Study 2: Lööf et al (2019),[19] Neurodevelopment Difficulties in Children with Idiopathic Clubfoot.
– Study 3: Lööf et al (2018),[20] Neurodevelopmental Difficulties Negatively Affect Health-related Quality of Life (HRQoL) in Children with Idiopathic Clubfoot

Patients and methods

Below is an overview of the methods used in the studies. Detailed information can be found in the individual papers. The Regional Ethics Committee in Stockholm approved all studies and the parents gave their informed consent for participation.

Study 1

In study 1, gross motor skills were assessed in 47 children with IC and 28 children without the deformity were recruited as controls (Table 1). The children performed the following motor tasks: running, walking, toe and heel walking, one-leg stance and hopping. The tasks were evaluated using the Clubfoot Assessment Protocol (CAP). In addition, initial clubfoot severity was assessed before treatment with the Dimeglio classification scale[21] and the passive range of movement of the foot at the time of the assessment were documented. On the CAP, each leg is rated individually for each task on a five-point scale from 0 (cannot/unable) to 4 (within normal).[22] We defined a score of 2 or below as deviant. Five blinded assessors rated the children individually from videotapes and the median scores were included in the statistical analysis.

Studies 2 and 3

In studies 2 and 3, NDDs and HRQoL were evaluated in four annual cohorts of children with IC from the counties of Stockholm and Skåne, Sweden. A total of 106 children with IC and 109 schoolchildren (control) in the same county areas were included (Table 1). In study 2, the children's parents answered the Five to Fifteen (FTF) questionnaire and Roye's Disease-Specific Instrument (DSI). The FTF has been developed to assess developmental and behavioural difficulties related to NDDs. The instrument entails 181 items representing eight domains (motor skills, executive functions, perception, memory, language, learning, social skills and emotional/behavioural problems) to be answered on a three-level score.[23] The DSI evaluates the parent's perspective of the clubfoot treatment outcome regarding function and satisfaction.[24] On the DSI, ten items are rated, also including questions about pain and appearance, and a total score can be calculated. In study 3, the children themselves answered the generic HRQoL questionnaire EuroQol-5D-youth (EQ-5D-Y) comprising of five dimensions ('mobility (walking about)', 'looking after myself', 'doing usual activities', 'having pain or discomfort' and 'feeling worried, sad or unhappy') as well as their perceived overall health status on a visual analogue scale (VAS).[25]

Table 1 Group characteristics of the participants within the studies

	n	Mean age, yrs (SD)	Boys % (n)	Bilateral IC % (n)
Study 1				
IC	47	5.4 (0.5)	74 (35)	47 (22)
Control	28	5.5 (0.6)	64 (18)	NA
Studies 2 and 3				
IC	106	9.4 (0.6)	73 (77)	46 (49)
Control	109	9.5 (0.6)	73 (79)	NA

IC, idiopathic clubfoot; NA, not applicable

Results

Gross motor skills

In study 1, significant lower CAP scores and a high percentage of gross motor deviations were found in children with IC compared with the control group ($p < 0.05$). This was especially prominent in the items 'one-leg stance' and 'hop' in which 87% and 85%, respectively, of the children with IC showed deficits (Table 2). Conversely, the controls demonstrated deviations of 32% and 43%, respectively, for the same items. In accordance with the CAP instrument, most children were still able to perform the motor tasks but did this with an impaired movement quality. For example, in the item 'one-leg hop' the child might have problems getting started, maintaining balance, keeping a straight line or with unregulated hopping strides.

A key finding in the study was that gross motor deviations were generally poorly correlated ($r_s \leq 0.4$) with the initial clubfoot severity and the passive range of movement of the foot at the time of the assessment (Table 2). This indicated that other reasons than musculoskeletal deficits alone, such as cognition and perception factors, may explain the high occurrence of gross motor deviations in this population.

NDDs

The results from study 2 showed that children with IC were reported with more NDDs on the FTF compared with the control group (Fig. 1). This was evident on a group level on the total FTF questionnaire, as well as in the following specific domains: 'motor skills', 'perception' and 'language' ($p \leq 0.04$; Fig. 1). In single items, 26 of the items

Table 2 Percentage of gross motor deviations in the study groups as well as the correlations between the motor tasks and the initial clubfoot severity (Dimeglio score) and passive foot movement at the time of the assessment of the idiopathic clubfoot group

	Percentage IC (n), (n = 47)*	Percentage control (n), (n = 28)	Correlations related to the gross motor tasks in the IC group†		
			Dimeglio score (n = 68)	DF (n = 69)	PF (n = 64)
Running	26 (12)	0 (0)	-0.22	0.40	0.16
Walking	26 (12)	0 (0)	-0.32	0.43	0.22
Toe walking	57 (27)	0 (0)	-0.31	0.06	0.28
Heel walking	64 (30)	4 (1)	-0.45	0.49	-0.05
One-leg stance	87 (41)	32 (9)	-0.24	0.13	0.12
One-leg hop	85 (40)	43 (12)	-0.11	0.13	0.13

*In this paper the inferior/worse performing legs in bilateral cases are presented, whereas the original paper presents all legs, including also the contralateral leg in unilateral IC[18]

†Correlations using the Spearman's rho; poor correlations defined as ≤ 0.04

IC, idiopathic clubfoot; DF, dorsal flexion; PF, plantar flexion

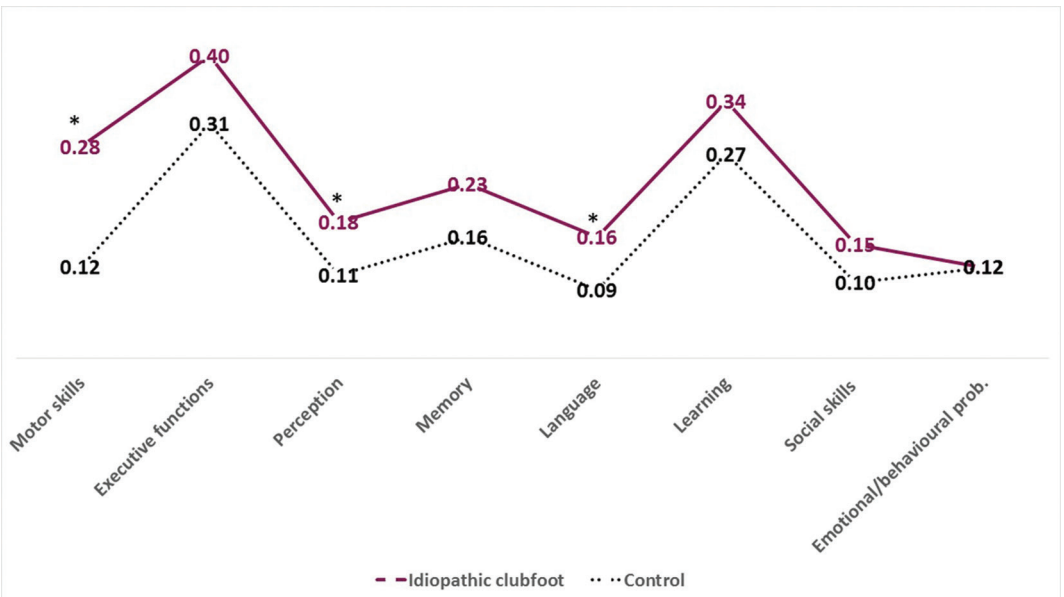

Fig. 1 Mean profiles of children with idiopathic clubfoot and the control group on the Five to Fifteen (FTF) questionnaire. Higher numbers indicate greater difficulties. Significant differences were found between the two groups on the total FTF as well as the domains with * ($p < 0.05$). The figure has been slightly modified from the original figure.[19]

were reported with more difficulties in the IC group. Interestingly, most of these items did not include musculoskeletal issues or the feet. On the contrary, the items consisted of concerns regarding areas such as social skills, difficulties handling small objects, writing and memory. Examples of this include the items 'difficulty behaving as expected by peers' and 'difficulty following and comprehending stories read aloud' that were reported to a greater extent in the IC group. The full list is detailed in Lööf et al.[19]

To define clinically relevant cases, children reported above the 90th percentile (with regards to the control group) of at least two domains on the FTF were identified in the IC group. This definition was in contrast to the guidelines of the FTF developer, who suggested that children above the 90th percentile in only one domain might be at risk of developmental disorders.[26] However, because the domain of 'motor skills' may be significantly affected due to the clubfoot diagnosis, we required two domains above the 90th percentile to consider a child at a clinically relevant risk. With this definition, 31% (n = 33) of children in the IC group were defined as clinically relevant cases with NDDs, i.e. at risk of developmental disorders. In this subgroup, the level of parental satisfaction of the clubfoot treatment outcome was found to be lower (p = 0.04) on Royes DSI (mean = 78 SD 19) than in those with only IC (mean = 86 SD 13).

HRQoL

In study 3, the findings revealed a similar high rating of overall health status in the IC (mean = 89 SD 14) and the control groups (mean = 90 SD 12) despite more problems being reported, primarily regarding pain and discomfort of children with IC. However, when considering the high prevalence of NDDs in the group of children with IC, we found interesting results. The subgroup of children with IC and clinically relevant cases of NDDs reported a lower overall health status (mean = 83 SD 17). In addition, only 30% of these children reported full health (no problems on the EQ-5D-Y dimensions), which can be compared with 59% of the children with IC only and 71% of the controls. Moreover, four of the five dimensions on the EQ-5D-Y showed significant differences between the two subgroups of children with IC (with and without NDDs) and the control group (Fig. 2). Thus, the coexistence of NDDs appears to be related to HRQoL problems in IC.

Discussion

The findings from the three studies suggest that IC may be more than an isolated structural deformity of the foot and lower leg in a considerable proportion of the children. Indeed, we found indications of additional challenges such as NDDs in this population. Although similar indications have been noted in previous publications, it has not been investigated to this extent before. Thus, the current findings are novel within the field of clubfoot and warrant further research in order to optimize treatment outcomes for children with IC.

In a study about childhood stroke, Max et al[14] found that 54% of boys with clubfoot demonstrated ADHD or

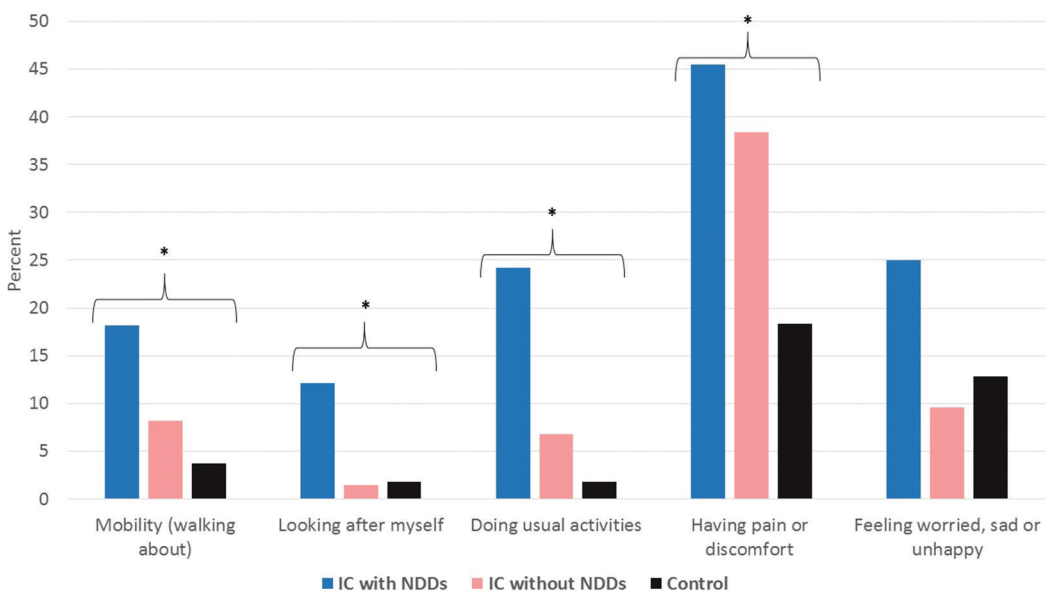

Fig. 2 The percentage of reported problems (some or a lot of problems) on the EuroQol-5D-youth of the children with idiopathic clubfoot (IC) with and without neurodevelopmental difficulties (NDDs) and the control group; * indicates significant differences between the three groups (p < 0.05). The figure has been modified from the original figure.[20]

traits of ADHD; astonishing findings that were never followed-up by the clubfoot research community. Our findings suggested that nine-year old children with IC demonstrated extended difficulties in areas such as language, attention and perception. Moreover, one-third of the children with IC were defined as being at risk of developmental disorders. This may also be an explanation of the lower movement quality found in children with IC, and that those motor deviations were poorly associated with passive range of movement of the clubfoot and initial severity. In line with this, in a large multicentre clinical study, Aulie et al[27] observed that no significant differences existed in motor abilities between children who had been treated with the Ponseti method or surgical management at nine years of age. The similarities between the groups were somewhat surprising to the authors since fewer children in the Ponseti-treated group had received major surgery. Therefore, the authors proposed that other factors could play a role in this, giving the example of a one-leg stand that also includes sensory and cognitive processes. Taken all together, there appears to be growing evidence of additional challenges should be considered in children with IC.

Clinical implications

In the treatment of IC, these additional challenges could considerably affect the treatment outcome. A notion supported by the finding that parents of children with both IC and NDDs reported a lower level of function and satisfaction on the Royes DSI (study 2). Several explanations for this can be considered. First, it might be possible that children with both IC and NDDs represent a different subgroup and/or a more severe clubfoot deformity. Secondly, children with this combination could be more difficult to treat for various reasons. For example, sleeping problems and sensations related to the skin (e.g. itching) have been reported to a higher extent in children with NDDs,[28,29] which may interfere with brace adherence and treatment outcome. Thirdly, parents of children with both clubfoot and NDDs may be harder to please. The potentially more prominent deformity of clubfoot could overshadow problems such as clumsiness and perception deficits, problems that might, in reality, be more associated with NDDs. Thus, NDDs might not be properly identified and the child and family could be left without treatment and support in such matters. Finally, parents themselves might experience difficulties, considering the heredity of neurodevelopmental disorders,[30] thereby introducing potential complications in managing the long and demanding bracing period in the treatment of clubfoot.

In a clinical setting, these additional challenges should also be considered with regard to pain management and in cases of surgery due to relapse. Pain was reported to a high extent in children with IC in our third study, both those with and without NDDs, highlighting the importance of pain management such as education about exercising and wearing proper shoes. Regarding the occurrence of additional NDDs, pain might also be related to those difficulties that require knowledge of such challenges. In cases of surgery, such as tibialis anterior tendon transfer, knowledge of this could be even more essential. First, as previously mentioned, in cases of motor deficits, a thorough motor assessment needs to be performed to determine whether it really is primarily a musculoskeletal problem for the individual child. It is possible that parents will seek medical advice if they are dissatisfied with the clubfoot treatment when the problem may in fact be more related to NDDs. Secondly, clinicians should consider whether the child and family will be able to handle the surgery, casting and bracing period. In cases of combined IC and NDDs, families may need additional support in order to be able to handle such an intervention. In some cases, it might be better to wait until the child and the family have the capacity and the support to undergo surgery. Finally, when a child with combined IC and NDDs has undergone surgery, he or she might need additional training in order to handle the new function of the muscle. To address such matters, a broad multidisciplinary approach that also includes health professionals such as psychologists, neurologists and physio- and occupational therapists in the treatment and follow-up of clubfoot would be beneficial.

The future

Our findings, combined with the indications in the aforementioned studies, may well have an impact in the search for possible shared genetic and/or environmental markers. In the search for a genetic aetiology of clubfoot, genetic data in NDDs may be of relevance also in clubfoot research. In the future, knowledge of clubfoot genetics could help clinicians inform individuals with clubfoot and their families about genetic aspects, as well as tailored treatment strategies and early identification of associated challenges. Moreover, future research ought to consider such challenges in longitudinal studies, for example by investigating predictive factors.

Conclusion

Our findings suggest the occurrence of additional challenges, such as NDDs, in a substantial proportion of children with IC that are beyond the structural musculoskeletal deformity, which may affect gross motor skills, clubfoot treatment outcome and HRQoL. Thus, clinicians working with this patient group should be aware of these

challenges and ought to consider thorough motor assessments and screening for NDDs. Furthermore, because of such difficulties, a multidisciplinary approach would possibly advance the care of children with IC – and their families.

Received 06 May 2019; accepted 21 May 2019.

COMPLIANCE WITH ETHICAL STANDARDS

FUNDING STATEMENT

No benefits in any form have been received or will be received from a commercial party related directly or indirectly to the subject of this article.

OA LICENCE TEXT

This article is distributed under the terms of the Creative Commons Attribution-Non Commercial 4.0 International (CC BY-NC 4.0) licence (https://creativecommons.org/licenses/by-nc/4.0/) which permits non-commercial use, reproduction and distribution of the work without further permission provided the original work is attributed.

ETHICAL STATEMENT

Ethical approval: All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards.

Informed consent: The parents in the studies gave their informed consent for participation.

ICMJE CONFLICT OF INTEREST STATEMENT

The author declares no conflict of interest.

REFERENCES

1. **van Bosse HJ.** Ponseti treatment for clubfeet: an international perspective. *Curr Opin Pediatr* 2011;23:41-45.

2. **Jowett CR, Morcuende JA, Ramachandran M.** Management of congenital talipes equinovarus using the Ponseti method: a systematic review. *J Bone Joint Surg [Br]* 2011;93-B:1160-1164.

3. **Sætersdal C, Fevang JM, Bjørlykke JA, Engesæter LB.** Ponseti method compared to previous treatment of clubfoot in Norway. A multicenter study of 205 children followed for 8-11 years. *J Child Orthop* 2016;10:445-452.

4. **Jeans KA, Karol LA, Erdman AL, Stevens WR Jr.** Functional outcomes following treatment for clubfoot: ten-year follow-up. *J Bone Joint Surg [Am]* 2018;100:2015-2023.

5. **Zionts LE, Ebramzadeh E, Morgan RD, Sangiorgio SN.** Sixty years on: ponseti method for clubfoot treatment produces high satisfaction despite inherent tendency to relapse. *J Bone Joint Surg [Am]* 2018;100:721-728.

6. **Sangiorgio SN, Ebramzadeh E, Morgan RD, Zionts LE.** The timing and relevance of relapsed deformity in patients with idiopathic clubfoot. *J Am Acad Orthop Surg* 2017;25:536-545.

7. **Desai L, Oprescu F, DiMeo A, Morcuende JA.** Bracing in the treatment of children with clubfoot: past, present, and future. *Iowa Orthop J* 2010;30:15-23.

8. **Seegmiller L, Burmeister R, Paulsen-Miller M, Morcuende J.** Bracing in Ponseti clubfoot treatment: improving parental adherence through an innovative health education intervention. *Orthop Nurs* 2016;35:92-97.

9. **Sangiorgio SN, Ho NC, Morgan RD, Ebramzadeh E, Zionts LE.** The objective measurement of brace-use adherence in the treatment of idiopathic clubfoot. *J Bone Joint Surg [Am]* 2016;98:1598-1605.

10. **Dobbs MB, Gurnett CA.** Genetics of clubfoot. *J Pediatr Orthop B* 2012;21:7-9.

11. **Bacino CA, Hecht JT.** Etiopathogenesis of equinovarus foot malformations. *Eur J Med Genet* 2014;57:473-479.

12. **Basit S, Khoshhal KI.** Genetics of clubfoot; recent progress and future perspectives. *Eur J Med Genet* 2018;61:107-113.

13. **Pavone V, Chisari E, Vescio A, et al.** The etiology of idiopathic congenital talipes equinovarus: a systematic review. *J Orthop Surg Res* 2018;13:206.

14. **Max JE, Mathews K, Manes FF, et al.** Attention deficit hyperactivity disorder and neurocognitive correlates after childhood stroke. *J Int Neuropsychol Soc* 2003;9:815-829.

15. **Garcia NL, McMulkin ML, Tompkins BJ, et al.** Gross motor development in babies with treated idiopathic clubfoot. *Pediatr Phys Ther* 2011;23:347-352.

16. **Andriesse H, Westbom L, Hägglund G.** Motor ability in children treated for idiopathic clubfoot. A controlled pilot study. *BMC Pediatr* 2009;9:78.

17. **Engström P, Van't Hooft I, Tedroff K.** Neuropsychiatric symptoms and problems among children with idiopathic toe-walking. *J Pediatr Orthop* 2012;32:848-852.

18. **Lööf E, Andriesse H, André M, et al.** Gross motor skills in children with idiopathic clubfoot and the association between gross motor skills, foot involvement, gait, and foot motion. *J Pediatr Orthop* 2017;1. (Epub ahead of print)

19. **Lööf E, Andriesse H, Broström EW, André M, Bölte S.** Neurodevelopmental difficulties in children with idiopathic clubfoot. *Dev Med Child Neurol* 2019;61:98-104.

20. **Lööf E, Andriesse H, Broström EW, et al.** Neurodevelopmental difficulties negatively affect health-related quality of life in children with idiopathic clubfoot. *Acta Paediatr* 2018. (Epub ahead of print)

21. **Diméglio A, Bensahel H, Souchet P, Mazeau P, Bonnet F.** Classification of clubfoot. *J Pediatr Orthop B* 1995;4:129-136.

22. **Andriesse H, Hägglund G, Jarnlo GB.** The clubfoot assessment protocol (CAP); description and reliability of a structured multi-level instrument for follow-up. *BMC Musculoskelet Disord* 2005;6:40.

23. **Kadesjö B, Janols LO, Korkman M, et al.** The FTF (Five to Fifteen): the development of a parent questionnaire for the assessment of ADHD and comorbid conditions. *Eur Child Adolesc Psychiatry* 2004;13(suppl 3):3-13.

24. **Roye BD, Vitale MG, Gelijns AC, Roye DP Jr.** Patient-based outcomes after clubfoot surgery. *J Pediatr Orthop* 2001;21:42-49.

25. **Wille N, Badia X, Bonsel G, et al.** Development of the EQ-5D-Y: a child-friendly version of the EQ-5D. *Qual Life Res* 2010;19:875-886.

26. **Korkman M, Jaakkola M, Ahlroth A, Pesonen AE, Turunen MM.** Screening of developmental disorders in five-year-olds using the FTF (Five to Fifteen) questionnaire: a validation study. *Eur Child Adolesc Psychiatry* 2004;13(suppl 3):31-38.

27. **Aulie VS, Halvorsen VB, Brox JI.** Motor abilities in 182 children treated for idiopathic clubfoot: a comparison between the traditional and the Ponseti method and controls. *J Child Orthop* 2018;12:383-389.

28. **Becker SP, Sidol CA, Van Dyk TR, Epstein JN, Beebe DW.** Intraindividual variability of sleep/wake patterns in relation to child and adolescent functioning: A systematic review. *Sleep Med Rev* 2017;34:94-121.

29. **de Schipper E, Lundequist A, Wilteus AL, et al.** A comprehensive scoping review of ability and disability in ADHD using the International Classification of Functioning, Disability and Health-Children and Youth Version (ICF-CY). *Eur Child Adolesc Psychiatry* 2015;24:859-872.

30. American Psychiatric Association. Neurodevelopmental Disorders. In *Diagnostic and Statistical Manual of Mental Disorders* (5th edition). Washington D.C.: 2013. http://dx.doi.org/10.1176/appi.books.9780890425596.dsm01. Accessed 1 April 2019.