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Pathological trunk motion during walking in children with Amyoplasia: Is it caused by muscular weakness or joint contractures?

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ABSTRACT

The aim was to investigate the causes for pathological trunk movements during gait in children with Amyoplasia. Eighteen children with Amyoplasia were compared with 18 typically developed children. Three-dimensional motions of pelvis, thorax and spine during gait were analyzed. Excessive trunk movements were defined as being above 4 standard deviations of those of typically developed children. Clinical examination of active strength and passive range of motion of the hip, knee and ankle joints were correlated to the parameter that showed the greatest prevalence of pathological trunk motion.

The greatest prevalence of 56% was seen for thorax obliquity range during walking. The spine angles showed the lowest deviations from typically developed children. Significant correlations ($p < 0.001$) between thorax obliquity range and clinical parameters were found for passive hip extension, hip flexion, hip abduction and active hip extension, hip flexion and ankle dorsiflexion strength. The highest correlation coefficients were found for passive hip flexion and active hip flexion strength of rho = −0.73 and rho = −0.69 respectively.

Excessive thorax obliquity during gait in children with Amyoplasia could be mainly caused by reduced strength and mobility of the hip. Therefore both mobility and strength of the hip are equally important and should be increased in the therapy to improve gait in children with Amyoplasia.

1. Introduction

Arthrogryposis multiplex congenita (AMC) is a heterogeneous condition which is characterized by multiple congenital contractures and muscular deficits in multiple body areas (Hall, 1997). The incidence of this syndrome was estimated between 1/3000 live births (Hall, 1997). It is believed that AMC is a result of fetal akinesia due to a number of different causes (Hall, 1997). The term 'Amyoplasia' refers to the most common arthrogrypotic syndrome that accounts for one-third of all patients (Hall, 1997). Amyoplasia is a condition characterized by a generalized lack in the newborn of muscular development

Abbreviations: AMC, Arthrogryposis Multiplex Congenita; ADN, Amyoplasia with natural Duchenne gait; ADE, Amyoplasia with excessive Duchenne gait; TD, Typically developed children; TOR, Thorax obliquity range.

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and growth with contracture and deformity at most joints; it is typically symmetrical and involves all four extremities with some variation (Bevan et al., 2007). In detail they have clubfeet which are associated with plantar flexor muscle weakness (Hoffer, Swank, Eastman, Clark, & Teitge, 1983), knee flexion contractures the hips are both flexed and external rotated. The shoulders are typically adducted and internally rotated, the elbows are extended with forearm pronation, wrists and fingers are flexed (Bevan et al., 2007). Children with Amyoplasia show maximum deformity at birth (Bevan et al., 2007). To increase their range of motion and to obtain a functional position of the joint, a combination of stretching, casting and often orthopedic surgeries are necessary (Staheli, 1998).

About 78% of patients with Amyoplasia are community level ambulators (Hoffer et al., 1983). However ambulation might require increased movements of the trunk to compensate for the contractures and weaknesses of the hip, knee and ankle joints. In children and adolescents with AMC increased lateral trunk sway, and rotation as well as pelvic lateral elevation, anterior tilt and transversal rotation were observed (Eriksson, Gutierrez-Farewik, Broström, & Bartonek, 2010). In particular the increased lateral trunk sway, also called "Duchenne gait" pattern (Duchenne, 1876) is most noticeable in public and lead to increased energy consumption (Duffels, Hill, Cosgrove, Corry, & Graham, 1996) and in consequence reduced walking distance. In addition Duchenne gait might increase the load on the spinal column (Kumar, 2004). The main reason for Duchenne gait found in the literature is abductor weakness (Duffels et al., 1996; Metaxiotis, Accles, Siebel, & Doederlein, 2000; Krautwurst et al., 2013). Another reason that has been speculated was, that lateral trunk lean assists foot clearance when either hip or knee flexion or ankle dorsiflexion are inadequate (Perry, 2010), which is quite common in Amyoplasia. Most therapeutic interventions have focused on the contractures. However, it has been shown in Amyoplasia that for motor function, muscle strength was more important for mobility than the passive range of motion (Kroksmark, Kimber, Jerre, Beckung, & Tulinius, 2006). Knowing the exact underlying reason for excessive trunk movements is important to find the best therapy to improve gait in patients with Amyoplasia.

Therefore the aim of this study is to determine increased trunk movements during gait in patients with Amyoplasia and determine its relation to clinical parameters of strength and passive range of motion in the hip, knee and ankle joints.

2. Materials and methods

2.1. Participants

In a retrospective cross-sectional study 18 children with Amyoplasia were included. Mean age was 8.8 (SD = 2.6) [4–12] years, mean bodyweight 29 (SD = 11) kg, mean body height 131 (SD = 18) cm, mean BMI 16 (SD = 3) kg/m^2, 12/18 were males. Eighteen typically developed children (TD) were measured for comparison. Mean age was 9.4 (SD = 2.3) [5–12] years, mean bodyweight 32 (SD = 10) kg, mean body height 139 (SD = 16) cm, mean BMI 16 (SD = 2) kg/m^2, 10/18 were males. All participants provided written consent, as approved by the local ethics committee. Patients had to be pain free and able to walk barefoot without assistance. Although few children used orthotics on daily basis, all gait studies were performed with the children ambulating barefoot. Exclusion criteria were spinal deformities and previous spinal surgeries, untreated hip dislocations and orthopedic surgeries within the last 24 months. Previous surgical procedures on the patients with Amyoplasia were listed in the following: 17/18 participants had clubfeet, of those, 10/17 were surgically corrected, 7/17 had Ponseti treatment following Achilles tendon tenotomies. 5/18 patients had surgeries to improve knee extension (3/5 supracondylar extension osteotomies, 1/5 knee flexor lengthening, 1/5 ventral epiphysiodesis), 5/18 patients had open hip reposition along with derotation and varisation osteotomies of the proximal femur combined with Pemberton pelvic osteotomy and psoas and adductor tenotomies.

2.2. Data collection

All participants underwent gait analysis with an 8 Camera system (Vicon MX, Oxford, UK) followed by a standardized clinical examination protocol listed in Table 1. The Vicon "Plug-in-Gait" marker set and model was used to determine orientation angles of the thorax and pelvis with respect to the laboratory frame. In addition spine angles were defined as the orientation angles of the thorax with respect to the pelvis. The subjects were asked to walk at comfortable speed down the 12 m walkway. Five consistent walking trials were used for the analysis.

2.3. Data analysis

Trunk motion of pelvis thorax and spine was reported in all 3 planes (sagittal tilt, sideward lean and transverse rotation). Selected parameters for the analysis were mean sagittal tilt, range of frontal lean and range of transverse rotation over the whole gait cycle. Mean sagittal tilt in the frontal plane was chosen instead of the total range, since the range of sagittal motion in these patients is typically low (Eriksson et al., 2010) and adaptive anterior pelvic tilt over the whole gait cycle due to hip flexion contractures (Perry, 2010) might have more relevance in patients with Amyoplasia.

The prevalence of excessive trunk motion was defined as the number of patients being outside 4 times the standard deviation of TD children during the left and/or the right gait cycle. Since most of the patients showed excessive deviations of the frontal plane thorax obliquity (TOR) this parameter was presumed to be most important and was correlated with the ROM and strength data listed in Table 1. Since contractures and muscular deficits in Amyoplasia are typically symmetrical at