The effects of knee-ankle-foot orthoses in the treatment of Duchenne muscular dystrophy: review of the literature

Authors' objectives
To assess the effectiveness of knee-ankle-foot orthosis in the treatment of Duchenne muscular dystrophy.

Searching
MEDLINE (1966 to 1997) and CINAHL (1982 to 1997) were searched for the joint occurrence of the words 'rehabilitation', 'muscular dystrophy', 'braces', or 'orthoses', and 'locomotion', or 'ambulation' in the title, abstract and/or keywords (subject headings). References from relevant publications were examined and recent volumes of relevant non indexed journals were examined. Articles published in Dutch, English, French or German were eligible.

Study selection
Study designs of evaluations included in the review
Uncontrolled clinical trials, controlled clinical trials and case studies were eligible if they were complete in their information on study characteristics, treatment, and quantitative presentation of the effect outcome or incomplete in only one of these three study characteristics. Included studies were controlled or uncontrolled trials or case series. Articles were excluded if no data on patients or outcomes measures were given, only the technique of bracing was described, or only the development of a specific type of brace was described.

Specific interventions included in the review
Knee-ankle-foot orthosis (KAFO) or calipers were eligible. Four types of modified KAFO were included: strong long leg braces, double bars, knee lock and limited ankle movement; contoured proximal leather band, providing support through ischial seating, double uprights, drop-ring locks, spring loaded ankles with sheepskin heel inserts, slight knee flexion and modifications of this type; steel, long-leg, double upright with knee spring lock and adjustable ankle stop; and long leg orthoses. Controversions included various active and passive exercise regimes, night splints, the use of walking aids for some boys and the following operative procedures to allow fitting of the orthosis: ilio-tibial band or fascia lata release; tendo Achilles lengthening; tendo Achilles release/tenotomy; hip flexors release; rectus femoris release; sartorius release; posterior tibial tendon transfer; and tensor fasciae latae release. Most patients had an operation on the lower limbs most frequently on the Achilles tendon. Training period (where stated) ranged from 17 days to 56 days and duration of use of orthosis ranged from 14 hours/week to daytime.

Participants included in the review
Boys with Duchenne muscular dystrophy were eligible. The boys ranged in age from 6 to 17 years and included those who had KAFO provided when they were: wheelchair bound for between 1 and 18 months; no longer able to walk; unable to walk independently; unable to stand for more than half an hour daily; able to stand independently but had limited ability to walk; or were still ambulant.

Outcomes assessed in the review
Inclusion criteria were not defined in terms of outcomes. A variety of outcomes measures were assessed in the primary studies with most studies reporting effect on ambulation but remaining vague about the definition of 'ambulation'. Outcomes assessed in the review included: success defined as patient not completely wheelchair bound after periods of one to three years; independent walking (child able to stand without support and able to walk without assistance); assisted walking (child unable to maintain balance while standing and walking but can walk with someone to steady him); standing ability; and effect of treatment on contractures and scoliosis.

How were decisions on the relevance of primary studies made?
The authors do not state how the papers were selected for the review, or how many of the reviewers performed the selection.
Assessment of study quality
Validity was assessed using the following seven study characteristics as described by Beckerman and colleagues (see Other Publications of Related Interest no.1): inclusion and exclusion criteria; description of study population; sample size; treatment; quantitative presentation of the effect outcomes; side-effects; and duration of effect. Studies were scored from 0 to 2 (maximum) on each characteristic with no data presented scored as 0, incomplete presentation scored as 1, and complete presentation scored as 2. Comment was also made on other aspects of validity (publication bias, selection bias, and measurement bias). The authors do not state how the papers were assessed for validity, or how many of the reviewers performed the validity assessment.

Data extraction
The authors do not state how many of the reviewers performed the data extraction.

Tables reported in the review included the following information: author and year of publication; methodological characteristics; study design; inclusion and exclusion criteria; characteristics of participants; sample size; KAFO type; operative procedures; training period; duration of use; co-intervention; main clinical outcome measure; % success; and duration of effect. The authors calculated the percentage of success in each study. The percentage of success was calculated as the percentage of patients not completely wheelchair dependent at one and three years. Data were extracted assuming a worst-case scenario in which every patient was regarded as completely wheelchair bound at the time of latest follow-up described in the studies.

Methods of synthesis
How were the studies combined?
Median percentage success of treatment was calculated after one year, two years and three years and median for the means of independent walking, assisted walking, and standing ability estimated.

How were differences between studies investigated?
The authors do not state how differences between the studies were investigated.

Results of the review
Nine studies were included in the review (526 patients with 484 patients treated with KAFO), including 3 case series, 4 uncontrolled group studies, and 2 controlled group studies.

Methodological characteristics from thirty-five studies were presented in a table.

The diversity of outcome measures in terms of ambulation made it difficult to summarise and compare the outcome results of all studies. The studies differed in the information on the type of orthosis used, various operative procedures performed, training periods, duration of use of KAFO, and often poorly described interventions.

Methodological considerations.

Potential sources of bias included publication bias, patient selection bias, and measurement bias.

Median percentage of success of treatment after one year was 75.1% (range 58.6% to 92.8%), after two years 47.9% (range 22.7% to 85.7%) and after three years 24.3% (range 4.5% to 29%).

Median for the means of independent walking (7 studies) was 24 months (range 19.2 to 32.6).

Median for the means of assisted walking (3 studies) was 36.2 months (range 0 to 90).

Median for the means of standing ability (2 studies) was 50.5 months (range 31.5 to 58.6).

Contractures and scoliosis: adequate data extraction was impossible because of missing data or because the progression of contractures was expressed differently in the included studies.
Authors' conclusions
The scientific strength of the studies reviewed was poor. It seems that the use of knee-ankle-foot orthosis can prolong assisted walking and standing, but it is uncertain whether it can prolong functional walking. The boys that benefit most have a relatively low rate of deterioration, are capable of enduring an operation and are well motivated.

CRD commentary
The aims were stated and inclusion criteria defined in terms of study design, participants, and intervention. Outcomes were not defined a priori. Two databases were searched and studies in any of four languages were eligible. No attempt was made to locate unpublished material and as acknowledged by the authors, publication bias was a possibility. Methods used to select studies were not described. Study validity was systematically evaluated, though methods used were not described. Relevant details of the primary studies were presented in tabular format. Given the heterogeneity among studies in intervention, patient characteristics and outcomes, it is doubtful whether pooling the data to estimate medians was appropriate. Potential sources of heterogeneity among studies were mentioned in the discussion and included differences across studies in terms of orthosis type, surgery, training period and duration of intervention. In summarising results from the review, attention was not drawn to better sources of evidence and results were not considered separately for controlled and uncontrolled studies.

The authors' conclusion as to the poor quality of evidence was supported. In view of the poor quality of studies, any other conclusions must be interpreted with caution.

Implications of the review for practice and research
Practice: The authors state that the use of knee-ankle-foot orthoses can prolong assisted walking and standing, but it is uncertain whether they can prolong functional walking.

Research: The authors state that a well-designed quasi-experimental study (executed by an independent investigator) with special attention paid to the avoidance of selection bias, measurement bias and confounding bias, might clarify the effectiveness of KAFO.

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Other publications of related interest

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Record Status
This is a critical abstract of a systematic review that meets the criteria for inclusion on DARE. Each critical abstract contains a brief summary of the review methods, results and conclusions followed by a detailed critical assessment on the reliability of the review and the conclusions drawn.