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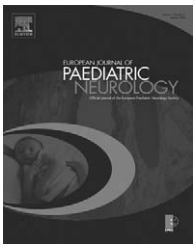

Original article

Knee–ankle–foot orthosis in children with duchenne muscular dystrophy: User views and adjustment

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ABSTRACT

Background: The use of knee ankle foot orthoses (KAFOs) to prolong independent mobility is a widely used rehabilitation strategy for children with Duchenne muscular dystrophy (DMD).
Aims: To explore views and adjustment of families with a child with Duchenne muscular dystrophy to the use of KAFOs.
Methods: interviews with families of children aged 8–18 years with DMD; questionnaires on psychiatric adjustment (SDQ for children; GHQ for parents).
Results: In total, 17 parents and 9 children took part. Families experienced the introduction of KAFOs as a signal for illness deterioration and a re-awakening of the feelings experienced at diagnosis. Nevertheless, the majority expressed a positive attitude and over two-thirds satisfaction with KAFOs use. High psychiatric risk was found in 2/17 children (12%; expected 10%) and 7/17 main carers (41%; expected 20–30%).
Conclusion: Most families were satisfied with KAFOs use, and its implementation was well tolerated especially by the children. However, mental distress was high in main carers who emphasized the importance of full preparation and support in this rehabilitation technique.

1. Introduction

Paediatric neuromuscular disorders encompass the breath of illness severity and functional impairment. Although no curative treatment exists, various supportive strategies have been shown to help improve outcome and provide solutions to overcome mechanical disabilities. However, their implementation makes demands on child and on family's psychosocial resources which are additional to those inherent to the underlying condition. Increasingly, medical interventions aim not just to improve medical problems but also to enhance function and the quality of child and family life. It is therefore important to assess the effects of any intervention on the child and family psychosocial adjustment.

Duchenne's muscular dystrophy (DMD) is an X-linked recessive condition, and with an incidence of 1 in every 3500 live male births, one of the most common genetic severe disorders of childhood, with more than 100 boys born in the

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United Kingdom alone every year. The defective protein, dystrophin which is indispensable for protecting muscle fibres from contraction induced muscle degeneration, results in progressive loss of muscle fibres and increase in weakness, and is accompanied by markedly elevation of serum creatine kinase. The first signs of muscle weakness occur between 1 and 3 years of age with difficulties in rising from the floor and running, followed by loss of independent ambulation at a mean age of 9.5 (range 6–12).[1]

The use of full leg KAFOs or knee ankle foot orthoses (KAFOs) has become part of the rehabilitation strategy of children with DMD. It is commonly instituted as a way of prolonging independent mobility at the time when independent ambulation becomes increasingly problematic because of the combination of proximal leg muscle weakness and Achilles tendon contractures. A review of eight reports documented treatment success at 1 year of 75%, falling to 48% at two and 24% at 3 years; the median successful independent walking achieved was 24, 36 months for assisted walking and 50 months for standing.[2]

The process of rehabilitation in KAFOs very often involves an orthopaedic surgical intervention (Achilles tendon release) and an intense physiotherapy training programme to allow the child to walk in these devices. KAFOs need often some readjustment to fit comfortably as they are fitted for most of the day, and their effect on mobility is not indefinite because of progression of weakness. Possible adverse psychological events, especially in older adolescent children, might be the realization of the progressive nature of the disorder. Conversely, the increase in independent mobility may help the development of optimism and improve psychosocial coping mechanisms.

An excess of emotional and behavioural problems have been described in children and young people with Duchenne muscular dystrophy[3–5] and high levels of stress in families.[6,7] The introduction of KAFOs represents a significant stage of illness progression and may be a risk period for the development of problems in psychiatric adjustment. In other paediatric chronic disorders, psychiatric problems in both children and parents can present at illness stages signifying illness deterioration.[8,9]

Webb[10] has explored strategies parents use to cope with their sons having DMD, and pointed out how they want to be heard and valued as experts on the disorders by medical and other professionals who interact with their sons, to participate proactively in their sons' lives and encourage other parents to do the same.

In this report, we describe parent and child views and satisfaction with the use of KAFO in children with muscular dystrophy, we document child and parent psychiatric adjustment and explore associations of satisfaction of KFO use.

2. Materials and methods

Participants and procedure: The study was conducted between May and October 2004. Children and adolescents over 6 years of age with DMD attending the Dubowitz Neuromuscular Centre at Hammersmith Hospital—a national centre in the United Kingdom for the assessment and treatment of childhood neuromuscular disorders—and their parents were approached for their willingness to participate. The centre is by far the largest in the United Kingdom and children attending represent a significant portion of the country's DMD population. Its rehabilitation strategies are similar to those in use in other centres in the country.

A research psychologist interviewed those who agreed and obtained questionnaire information. Clinical information was available from case notes and through discussion with clinicians and the Muscular Dystrophy Campaign Family Care Officer, who attended the clinics from which the children were recruited. The research had a qualitative or descriptive and a quantitative dimension.

The rehabilitation in KAFOs is offered to all DMD children at the end of their functional walking, with the exception of those boys with severe learning disability, unable to comply with the process, or with significant co-morbidity (i.e. dilated cardiomyopathy). Approximately, 90% of DMD boys followed up are offered the rehabilitation through KAFOs. The procedure followed in our centre has been described in detail.[11] Briefly, in order to fit KAFOs the large majority of DMD boys require transaction of the Achilles tendon, as by the end of the functional walking they have a significant equinous deformity of both feet. The surgical intervention is performed under general anaesthetic and followed by a period of rehabilitation of approximately 7–10 days, of which at least the first 3 days are inpatients because of the necessity for pain relief. The mean duration of walking in KAFOs following the procedure is 24 months. We have recently evaluated a large population of DMD children for risk factors for the development of scoliosis, and found that standing and walking in KAFOs was the most significant factor associated with protection from the development of scoliosis (Kinali et al., manuscript in preparation).

The qualitative/descriptive arm consisted of semi-structured interviews with children and main carer (normally mothers), seeking their views about the use of KAFOS. In order to obtain information about different stages in the process, a purposeful qualitative sample was chosen whereby interviews were conducted with children and families at three different phases: (a) current use; (b) past use; (c) where the use of KAFOS had been considered. I Interviews explored attitudes (such as satisfaction or lack of this) and experiences about practical use of KAFOS. It included questions on effects on the child's mobility, emotional and social life, and on what facilitated or hindered use.

The quantitative arm involved information about demographic data (age, parental situation, social class and geographical distance from clinic). Illness impairment was quantified with the Functional Disability Inventory or FDI, a questionnaire of documented validity, stability and sensitivity to change in general populations.[12] We documented attitudes towards calliper use through 6-point Likert scales indicating global satisfaction with or positive expectation towards their use, and degree of support received from hospital staff, family, friends or any other sources.

Standardized questionnaires were used to assess child and parent psychiatric adjustment. The Strengths and Difficulties Questionnaires (SDQ)[13] is a 25-item questionnaire that enquires about a child's symptoms in the previous 6 months.