Prosthetics and Orthotics International, 1992, 16, 64–68

Case study

A remarkable transport device for a fibrodysplasia ossificans progressiva patient

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Abstract

This article contains a case report and a general description of a patient with fibrodysplasia ossificans progressiva.

This is followed by a description of the design of a standing and transport device.

Introduction

The Academic Medical Centre (AMC) in Amsterdam is one of the largest hospitals in the Netherlands. A great deal of scientific research is done at the AMC and it also accommodates a training centre (University of Amsterdam).

The rehabilitation department of this hospital encompasses the following disciplines:

- physiatry;
- physiotherapy;
- occupational therapy;
- orthopaedic instrument making;
- social work.

Because of the research facilities of the AMC, many rare diseases are seen in the hospital and are often sent for treatment to the rehabilitation department.

One of these patients was suffering from fibrodysplasia ossificans progressiva (FOP). His recurrent decubitus ulcers and problems in moving and standing were the reasons for his referral. After analysing the specific problems of this patient, a special standing and transport device was designed by an occupational therapist and a physiatrist.

General description of FOP

Fibrodysplasia ossificans progressiva (FOP, formerly known as myositis ossificans progressiva) is a rare disorder in which there is progressive ectopic ossification and characteristic skeletal malformation (Connor and Evans, 1982; Rogers and Blair Geho, 1979; Heutzer *et al.*, 1977). It is an autosomal dominant disease, but it usually occurs as a fresh mutation.

It is likely that there is a paternal age effect related to occurrence. Because of the early onset of symptoms there is a lack of reports concerning offspring. The skeletal malformation consists of:

- abnormal big toes. These are mostly short, monophalangeal with valgus deviation;
- short thumbs, due to short metacarpals;
- short broad femoral necks;
- abnormal cervical vertebrae with small bodies, large pedicles and large spinous processes.

Formation of ectopic bone usually starts in early childhood in the neck of the dorsal paraspinal muscles. The initial manifestation is the appearance of tender soft tissue swellings, which may progress in non-tender ossified nodes. Only four to six months after the appearance of these swellings there is radiological evidence of ossification. Certain areas of connective tissue are especially prone to ossification. These areas include the connective tissue of the paraspinal muscles, the muscles in the limb girdle and the mastication muscles. Joint capsules, ligaments and plantar fasciae are also often involved. Calcification, affecting the shoulders, elbows, hips and knees is commonly bilateral. The heart, tongue,

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larynx, diaphragm, perineum, eye and sphincters remain unaffected. Involvement of the abdominal wall is uncommon. There is no involvement of the skin by ossification, but ulceration of the skin over a projecting spur of bone or due to pressure sores is an occasional feature. Other features can be precipitated by a number of factors such as trauma to the muscle, biopsy of the swellings, operations to excise ectopic bone, intramuscular injections, careless venipuncture and dental therapy. Episodes of inactivity can last for many years. The ectopic ossification is most marked before puberty but new swellings can still occur in the sixth and seventh decades of life.

The progression of disability is not correlated with the sex of the patient, the age or site of onset of ossification, the type or extent of skeletal malformation, nor is it affected by medical treatment.

Because of the severe reduction of mobility, most patients either require assistance in activities of daily living or are totally unable to undertake any activities at all. Management of the patient should concentrate on the avoidance of exacerbating factors, prevention and prompt treatment of pneumonia, the provision of adaptations in order to cope with the activities of daily living, and the prevention of decubitus ulcers.

Case report

The patient described in this case report is a 41 year old man suffering from severe FOP. He has had ossifications since the age of seven. He was born when his mother and father were 32 and 37 years old respectively. There was no consanguinity of the parents. No members of the family had malformations or diseases of connective tissues (including two older brothers and one older sister). The pregnancy and delivery were without complications. At birth he showed a bilateral hallux valgus, a microdactyly of the great toes (monophalangic) and of the thumbs (due to short first metacarpals). The first swellings and ossifications occurred when he was seven years old, at the thoracic and cervical spine. Since then the disease has progressed steadily.

At the time of writing the largest part of his body has become fixed in one rigid position. The following specific characteristics can be observed with this patient:

- the cervical spine is completely stiff, which causes difficulties with eye-hand co-ordination;
- the thoracic and lumbar spine are also in complete ankylosis with a slide anteflexion and left latroflexion. This position produces a tendency to fall sidewards. He uses a stick to prevent this;
- the legs are ankylosed in all joints. There is a slight flexion position of the left knee and hip, and less of the right knee and hip;
- the right arm is the most functional. There is a limited function, about 20 degrees in all directions of the shoulder, and a good function of the elbow, wrist and hand. He is able to eat, hold a book and handle the television by remote control with the use of his right hand.

In the left arm there is a good function of the wrist and hand. However, there is a severe limitation of the elbow and an ankylosis of the shoulder. Therefore it is impossible to use his left hand for anything else but holding his stick while standing.

There is a severe restriction of the jaws, resulting in eating problems and problems with dental care.

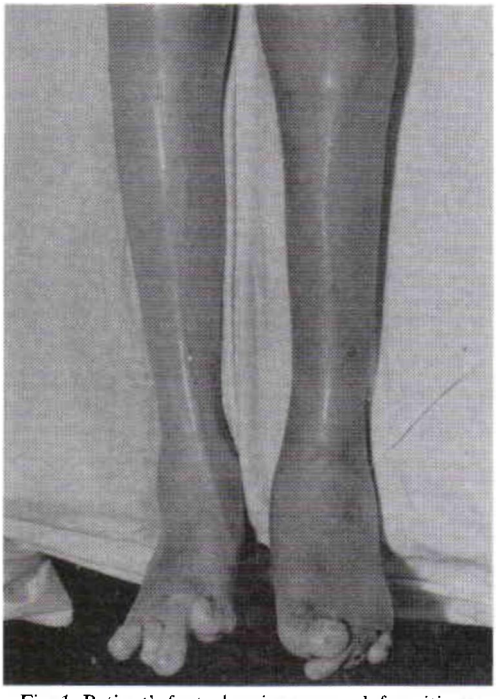

Fig. 1. Patient's feet, showing severe deformities.

The thoracic expansion is totally inhibited. Sufficient respiration is still possible, because his abdominal wall is not involved. He has not had pneumonia until now.

When he first presented, his major problems were recurrring pressure sores of both feet over a 14 year period, and the left thigh for a period of one year. After having provided him with a special anti-decubitus mattress (a ROHO mattress), the problems of his thigh were solved. Because of the severe deformity of his feet (Figs. 1 and 2) rehabilitation shoes were provided to achieve an optimal distribution of bodyweight over the whole of both foot soles and to give better balance while standing. With optimal fitting, however, the feet remained a problem. It appeared that his feet could not bear his bodyweight any more during the whole day. Because his only other alternative was lying in bed and because of his wish to move without help, a standing device was designed. In this standing device he should be able to drive his wheelchair and change position in order to relieve his feet.

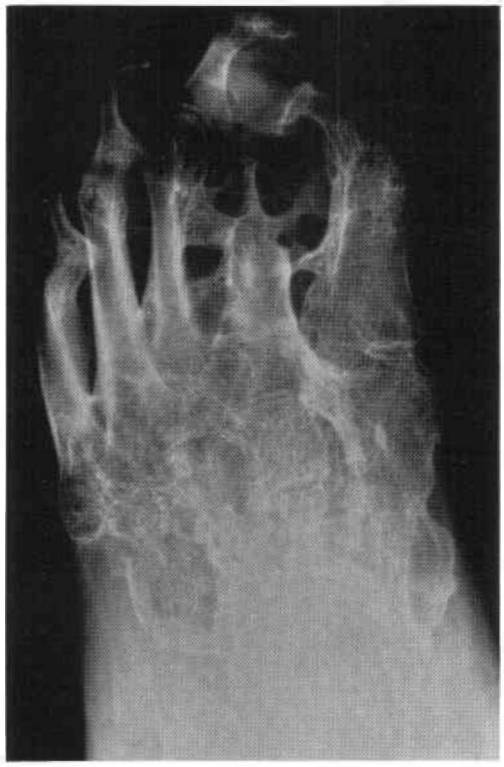

Fig. 2. X-ray left foot at the age of 40, showing severe ossification and osteolysis based on recurring ulcerations.

Design

Before starting to design the device a problem analysis was carried out, containing the following components:

the patient's physical level of function, as described above;
the medical goal of the device;
the user's wishes concerning the device;
specific safety comments.

Medical goals

To relieve the pressure on the feet, it is necessary to make it possible to change position from lying to standing in an easy way. To prevent decubitus ulcers elsewhere on his body, the body contours should be accurately followed. An optimal spread of pressure and a good air circulation (to prevent extra perspiration in a totally fitted device) were required.

User's wishes

The patient had the following wishes concerning the device:

— to rise from and sit down in his chair independently;

Fig. 3. Patient standing in his device. Later the position of the armrest and footboard were changed.

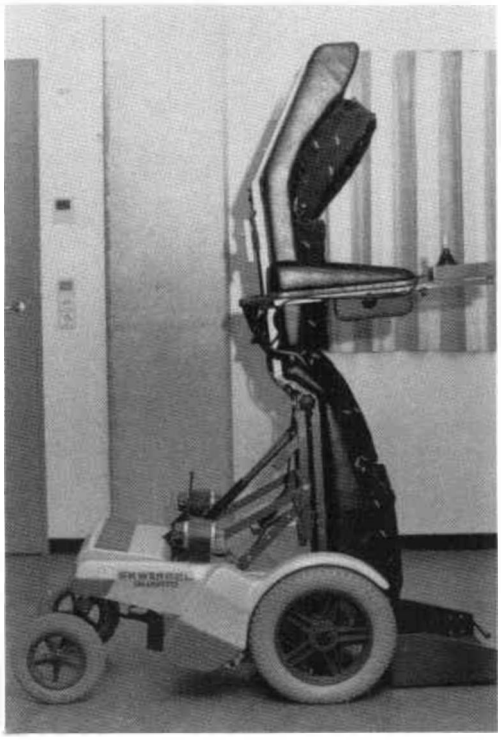

Fig. 4. Upright standing position in which he is able to leave and enter the device. Note the curves necessary to follow the patient's ankylosed posture.

— to move in and outdoors without help;
— to drive his wheelchair on slightly rough terrain, such as a garden or a camp site; this really was a great desire;
— to change from standing to lying and back in a very easy and independent way.

Specific safety comments

The patient has to drive in an almost upright standing position so that he can see where he is going. This means great stability is necessary for the undercarriage. It must be possible to use safety devices (such as belts) independently. The driver's control has to be visible as well as easy to operate with his right hand.

Practical development of the device

After completing the problem analysis, the manufacture of the device was commenced (Figs. 3—5). A children's wheelchair undercarriage (HUKA-Squirrel) was used as a base. This undercarriage has a standard feature high-low control, which was transformed into a rotating function. The undercarriage is designed to be very steady, because of the high-low function. It has a front wheel drive, which makes it suitable for use on rougher terrain. A square metal frame was fixed to the base. The frame was fitted approximately to the patient's body. Since the neck and the hips of the patient had a flexion position, two bends were made in the frame. After this, the frame was covered with foam. The top was made of a ROHO low-profile anti-decubitus mattress. As a footboard, a very thin metal carrier was used. The footboard came flat on the ground in the upright standing position. This made it possible for the patient to enter the chair without help.

After completing the building of the chair, the driver's control was placed in a good position. Safety aspects were as follows:

— the elbow-rests were extended from the trunk and were made tip-up;
— for outdoor-driving a safety belt was added.

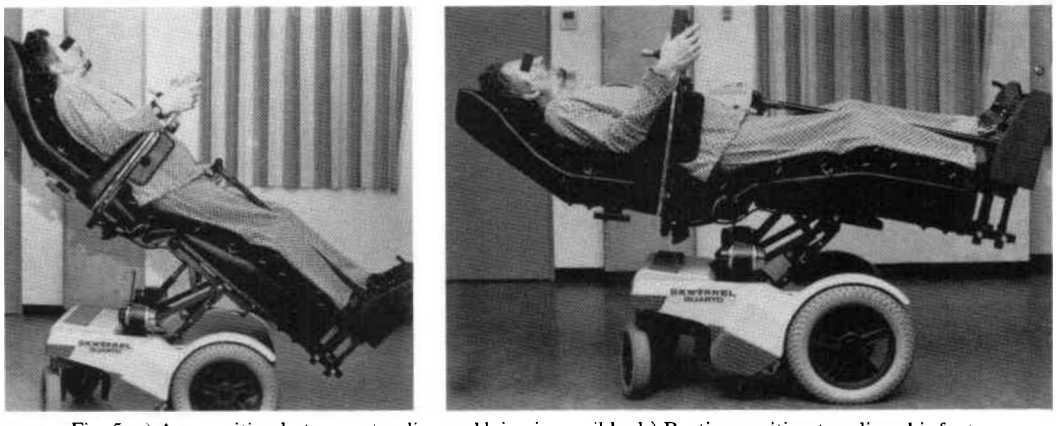

Fig. 5. a) Any position between standing and lying is possible. b) Resting position to relieve his feet.

When standing the left leg is standing more in anteflexion than the right leg. Therefore, the left foot was extended somewhat over the ridge of the footboard. To avoid stubbing his toe, a tip-up frame for the left foot was made. This frame could be handled independently by the patient with the help of his stick.

Evaluation of the user

The chair is regarded by the patient as very comfortable. His action radius has improved enormously, and he has the opportunity to go out on his own. Since he has had his chair no new decubitus ulcers have occurred, and the ones he had have healed.

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