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## BILATERAL AGENESIS OF UPPER EXTREMITY

JOSEPH L. FLEMING, M.D.\*

Every doctor is, to some extent, an expert in rehabilitation. The word simply means helping a person by many and various means to achieve maximum biological efficiency. No one group may rightfully lay exclusive claim to the word and, in fact, by its nature the rehabilitation process is necessarily a team project in which each specialty group plays its role under the coordination of the specialist in whose field the major disorder lies.

Optimism is the chief password in the rehabilitation process and many a forbidding sentinel of hopelessness will step aside when this magical word is invoked. In orthopaedic surgery we have found it a good rule when dealing with severe disruption of function, whether congenital, traumatic or infectious, to assume the best possible outcome and to treat vigorously on the assumption that some day the child may be able to walk. For example, if a child with severe meningomyelocele does not ever walk, it should not be because the orthopaedic surgeon has allowed the development of deformities of knees, hips and feet that prevent bracing and ambulation.

Looking backward on the case of Patricia K., it seems obvious that she would walk, use artificial arms, feed herself and attend school, but if it were not for the optimism bred by the despair of her mother, she would still be in her cart, writing and eating with her feet, and deprived of the advantages of association with other children in school.

### Figure 1 and 2

Standing with support before treatment.

Patricia K. was born in 1948 of normal parents and following a normal pregnancy. There was a complete agenesis of the arms although the scapulae and clavicles were normal. There were bilateral clubfeet, bilateral absence of the fibulae, and bilateral congenital dislocation of the hips. There was a moderate right dorsal scoliosis.

The hips were reduced by Dr. Gehring of Detroit and the feet were casted for a year until the parents discontinued treatment. At age 3, an attempt at bracing and

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ambulation failed, at least partially due to the severe foot deformities.

When first seen in 1953 she was four and a half years old. She was transported by her mother in a cart and it was becoming increasingly difficult for her mother to lift her. The chief reason for the parents bringing the child was to see if a way could be found for the child to walk or to gain some increased independence.

Patricia could be stood up but required a good deal of help from supporting hands. The arms were absent and in the glenoid fossa was a slightly protruding pad of thick fat. The musculature of the shoulder girdle was relatively good except for weak pectorals. There was a moderate right dorsal scoliosis of 27 degrees. Both hips were stable although x-ray revealed that the right hip was dysplastic and subluxated. Both knees were very unstable and dislocated anteriorly on extension. The fibulae were absent.

The feet had grossly defective musculature especially the anterior tibials and peroneals. There were semi-rigid equinovarus deformities of severe degree.

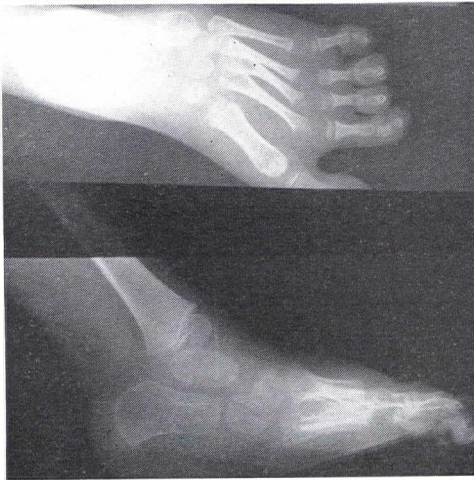


Figure 3

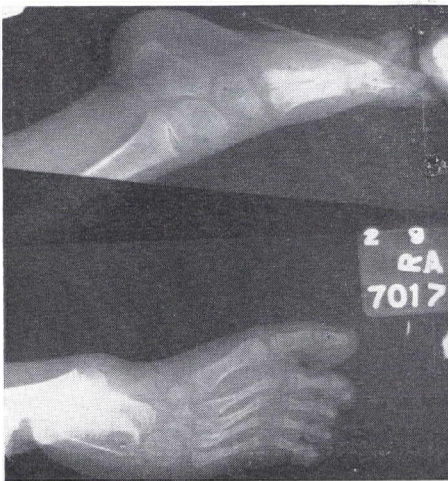


Figure 4

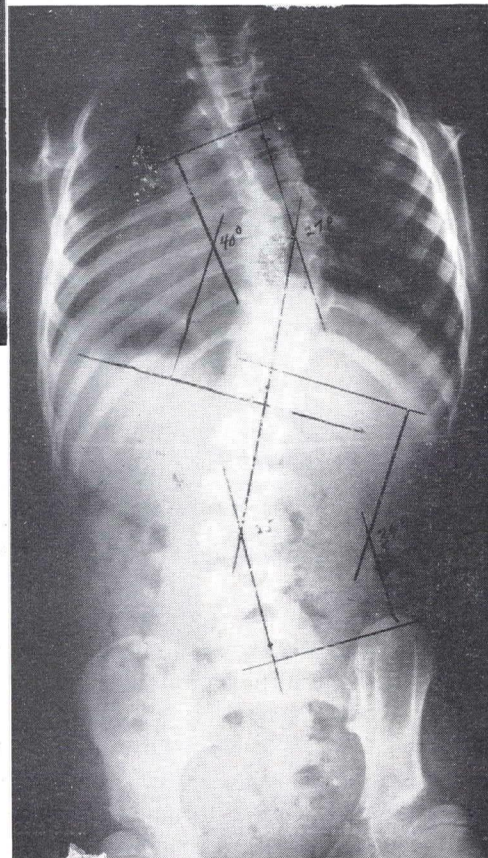


Figure 5

Figures 3, 4 and 5 — Congenital scoliosis and equinovarus foot deformities.

Figure 6-7-8

Skill in use of feet, age 4 years.

In spite of these deficiencies, she had developed an astounding skill with her feet. She was able to feed herself, write, color, comb her hair, etc. The hallux functioned much as does a thumb. Sitting down, with her shoes off, she had remarkable efficiency. She pushed herself about the floor very efficiently. All in all, an attractive and

charming sight in a four-year old but not so acceptable in a woman of fifty.

Psychological testing disclosed an I.Q. of 85 which was considered a minimal score.

Patricia was seen in consultation by many physicians from many specialities and opinion was unanimous and vociferous that nothing should be done to the child in order to make her walk that might endanger the use of her feet as hands. In addition, it appeared to everyone after seeing her that due to the multiple defects and deformities that independent walking was most unlikely, and the great fear that we would lose her abilities without gaining anything.

The parents however strongly supported the orthopaedic view that to walk, to attend school, was at least as important as the possible partial loss of prehensile pedism. The mother's words, "I can always feed her and comb her hair but I can't always lift and carry her," precipitated the decision to proceed with her rehabilitation.

First the feet were wedged with plaster casts, out of forefoot adduction. This required six weeks and was stopped short of full correction because of the fear of developing severe deformities which are usually present when the fibulae are absent. The feet then could be brought passively to the neutral position from their previous position of thirty degrees of forefoot adduction.

Next the feet were wedged out of the position of thirty degrees of equinus. This proceeded very slowly and consumed ten weeks. The feet could now be brought up to the neutral weight-bearing position. They were very stiff and loss of function was feared. The corrected position was maintained by passive stretching, night casts and active exercises while the Physiotherapy Department worked on developing the upright muscles — the quadriceps femoris, gluteus maximus and spine extensors — all of which had been of very poor quality due to disuse. A fracture of the tibia further prolonged this stage. After four months of intensive physiotherapy and a strenuous home exercise program, she was able to walk with help but by herself she could balance only for a few seconds. She had regained full use of her feet.

A Taylor spinal brace was fitted with a short length of broom stick attached by a swivel joint to its back. With this apparatus the parents could walk her about for long periods, gradually decreasing the amount of assistive balancing. She was taught

Figure 9-10  
Standing alone after treatment.

to use her head to replace the normal balancing function of the arms. After four weeks with the brace, ten months after her treatment began, she walked independently. The stability of the knees increased as the quadriceps were developed and tibial osteotomies, previously considered, were not indicated. She was trained to rise from a sitting position and also taught how to fall without injury.

About this time an Amputation Clinic was developing at this hospital under the direction of Dr. C. Leslie Mitchell, and previously unfittable upper extremity amputations was now being prescribed for. Patricia was trained in the use of a right shoulder disarticulation prosthesis and within a few weeks had mastered the fundamentals. Six months later a left prosthesis was prescribed and she has become proficient in its use as well. These prostheses are controlled by contralateral shoulder harnessing and ipsilateral thigh harnessing.

Figure 11, 12, 13, 14 — Skillful accomplishments, age 9 years.

Figure 11-12-13-14

Skillful accomplishments, age 9 years.

A partial list of her accomplishments are: 1. Feeds herself pre-cut food with adapted utensils. 2. Brushes teeth, washes face, combs hair. 3. Types with two hands. 4. She attends school and camp for handicapped children. 5. She is fully independent in school except for dressing and personal hygiene. 6. She is now included in children's activities at home. 7. She keeps her shoes on all day and is using her feet only occasionally and in private.

Many problems still lie ahead for this child. It is possible that the hips and knees and feet will deteriorate with use and so diminish her efficiency in walking. However, this would be equally true for her efficiency in using the feet as hands. She will need long continued and expensive care in the future with regard to her prostheses. She is now potentially employable and her increased independence has been gratifying to all.

This child was cared for under the Michigan Crippled Children's Commission program with the invaluable co-operation of Dr. Clarence Eisman, M.D. She was fitted with her prostheses by Dr. James Glessner, M.D. and Dr. Charles Long II, M.D. and was trained in their use by Miss Powell of Occupational Therapy. Mr. Tony Fillipis of Wright-Fillipis was the prosthetist.