

# Terminal Transverse Congenital Limb Deficiency of the Forearm

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## INTRODUCTION

Although many investigators have studied congenital anomalies of the upper limb, few have focused their attention on complete congenital limb defects. These patients share certain problems with those having traumatic amputations or surgical amputations for neoplastic or infectious disease. However, the patient with a congenital limb defect has additional problems and different needs. Proper attention to identification of these differences is critical in providing the best possible care.

## PURPOSE

The purposes of this study are: (1) to examine patients with terminal transverse congenital deficiency of the forearm, (2) to describe prosthetic care for these patients, and (3) to describe the attitudes of patients and parents toward prosthetic treatment.

## METHOD

Patient records for 2527 patients have been classified in the files of the University of Iowa Congenital Hand Project. A review of the records of all patients with congenital forearm amputations was made (Fig. 1) and a

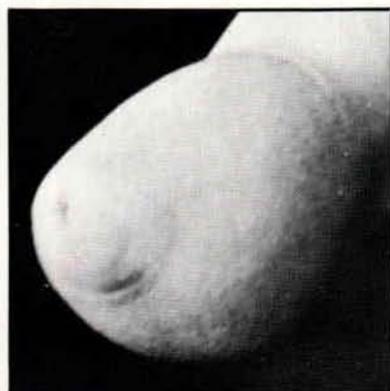


Fig. 1—Classic TTCLD Forearm Shurr, Cooper, Buckwalter & Blair Terminal Transverse Congenital Limb Deficiency of the Forearm

standardized list of questions established. All available patients were evaluated during an outpatient clinic visit. All information from both the patient and parent(s) was obtained by the senior author. Some patients in this series have been lost to follow-up and appear only in the incidence section of this report.

## REVIEW OF LITERATURE

Birch-Jensen (3) examined records of over four million patients to determine the incidence of the below elbow amputation. In this classic study (Fig. 2) a total of 161 patients were identified as congenital below elbow

## OCCURENCE OF CONGENITAL BELOW ELBOW AMPUTATIONS

	Shurr (1980)	Birch-Jensen (1949)	Aitken & Frantz (1955)	Aitken & O'Rahilly (1961)
Number of Patients	48	161	49	331
Male (%)	19 (40)	69 (43)	22 (45)	156 (47)
Female (%)	29 (60)	92 (57)	27 (55)	175 (53)
Left (%)	35 (69)	108 (67)	37 (76)	212 (64)
Right (%)	16 (31)	53 (33)	12 (24)	119 (36)

Fig. 2—The Slide of All Studies Shurr, Cooper, Buckwalter & Blair Terminal Transverse Congenital Limb Deficiency of the Forearm

amputees. 69 were male, 92 female; 108 occurred on the left and 53 occurred on the right. Aitken and Franz (1) reported a total of 49 patients; 22 males and 27 females, 37 lefts and 12 rights. In a series published by Aitken and O'Rahilly (2) a total of 331 cases were reviewed. Of these, 156 were male, 175 female; 212 were lefts and 119 were rights. The data in each of these studies agrees in terms of relative incidences, indicating a predominance of females and a left to right ratio of nearly 2:1.

## RESULTS

Forty-eight patients with below elbow amputations were identified (Fig. 2). These patients were placed into two groups: Group 1—unilateral below elbow congenital amputees, and Group 2—patients with associated anomalies, to include bilateral below elbow congenital amputation. There were four bilateral upper extremity amputees. There were 19 males and 29 females. To complete the series, one patient with a below elbow amputation also had a contralateral elbow disarticulation, making a total of 52 amputations in this series. Of 51 below elbow amputations, 35 were on the left and 16 were on the right.

Infants seen by the University of Iowa Department of Orthopaedics in recent years are fitted with a plastic below elbow socket, suspension strap, and a passive paddle, as early as age five months (Fig. 3). Physical

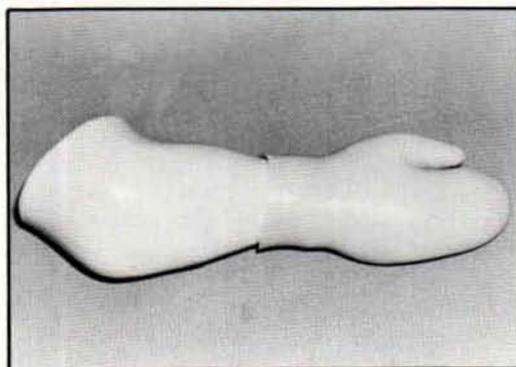


Fig. 3—B.E. with Paddle Shurr, Cooper, Buckwalter & Blair Terminal Transverse Congenital Limb Deficiency of the Forearm

therapists instruct the parents as to evaluation of proper fit, means of donning and doffing, how to assist the child in using the device, what to expect functionally, and how to check the skin for signs of an ill-fitting prosthesis or tight harness. Attention is given to the subject of prosthetic tolerance. It is recommended that unilateral below elbow amputees wear their prostheses all day from an early age on. Return visits are scheduled to follow the child and answer questions of concerned parents.

When the child outgrows the initial prosthesis, usually at the age of one and one-half to two years, a new socket is made and a split-hook Dorrance terminal device is introduced (Fig. 4). Care is taken to educate the parents to the body motions needed to power



Fig. 4—B.E. with Split Hook Shurr, Cooper, Buckwalter & Blair Terminal Transverse  
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the voluntary opening terminal device. The timing of the first split-hook prosthesis allows about one year for the family to become accustomed to the child's amputation and to the future use of a prosthetic hook. In America, we live with the negatives associated with the fictional pirate, "Captain Hook." Few families will accept a split hook for a prosthesis for their six-month-old child, even if it could be functional.

A successful wearer may be defined as a person who wears the prosthesis most of the waking hours. Using this definition, many successful wearers were fitted prior to age one. Parents of successful wearers expressed satisfaction in the aggressive, early fitting approach. Most are eager to talk to other parents and relate how quickly their child used both limbs, once the fitting of the below elbow prosthesis occurred. Parents also report that functional milestones are often delayed, including dressing independence and tying one's shoes. Most parents comment about the improved function of their child with the use of the device. This is difficult to measure objectively, since no controls exist and since a comparison with a normal limb would be unfair. However, successful wearers are not necessarily successful users and, as demonstrated by one farmer who wore his arm only for certain tasks, a successful user may not always be a successful wearer. Concerning

the appearance of the prosthesis, parents often describe it as "cold," "clunky," "ugly," or "noisy," but most of these same families admit that their child looks "naked without it."

Questions concerning deficiencies in the prosthetic device or hook indicated most successful wearers and their families feel the devices are adequate. Many refer to the day in the future when a prosthetic hand will be as practical and useful as a hook. In contrast, many teenage children, who frequently are concerned about their appearance, report discontinuing use of a prosthesis through the ages of 13 to 20, only to return to prosthetic use at a later age.

## DISCUSSION

An unsuccessful wearer seldom wears the prosthesis. The unsuccessful wearers can be categorized as those who were fitted after the age of five, and some after the age of 10. Drastic changes in wearing history appear to be rare. Charts reviewed from the 1940s commonly revealed references to late-childhood or even adolescent-age fitting as the recommendation of choice.

Children with both arm and leg deficiencies present a particularly interesting problem. Early lower limb fitting appeared to be

based on the chronology of motor skill development, but the philosophy of fitting upper extremity amputations was not based on the child's motor development.

We have adopted the philosophy of early fitting, recognizing that this approach results in successful wearers. In reviewing the children fit as adolescents, successful wearers were few. One patient felt her skills with just the elbow crease and normal hand were equal to her abilities with a prosthesis. Others feel that the prosthesis gives the appearance of a "handicapped person," and going without anything is more satisfying to their self image.

No patients have been fitted with myoelectrically controlled electric hands, although the Muenster design socket is the socket of choice in the adult below elbow wearer. For those adults who desire to lift heavy loads, such as farmers, more conventional socket and harnesses are used.

## CONCLUSIONS

The congenital below elbow terminal transverse amputation appears to be a distinct entity, well defined in its unilateral presentation. It occurs in our series more often in females (29 versus 19) and more often on the left (35 versus 16). Early, aggressive fitting of prostheses at about six months of age is well accepted by both parents and children. This approach yields a functional prosthesis, at a very young age, and appears to lead to successful adult wearers.

## REFERENCES

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