The Journal of the International Society for Prosthetics and Orthotics

Prosthetics and Orthotics International

Special Issue
The Limb Deficient Child

August 1991, Vol. 15, No. 2


3C1 – the new generation kn
from OTTO BOCK
Step into the future of a more activ
and zestful life
Stability and minimal weight provid
technology
Safety and dynamics through hydr
Modular system adaptability and le
adjustability
A great accomplishment supportin
progressive rehabilitation.
*Mauch SNS-Hydraulic
Otto Bock
ORTHOPÄDISCHE INDUSTRIE
Industriestraße · Postfach 12
D–3408 DUDERSTADT
© OTTO BOCK 05.1991

Prosthetics and Orthotics International

Prosthetics and Orthotics International is published three times yearly by the International Society for Prosthetics and Orthotics (ISPO), Borgervaenget 5,2100 Copenhagen Ø, Denmark, (Tel. (31) 20 72 60). Subscription rate is GBP55 per annum, single numbers GBP20. The journal is provided free to Members of ISPO. Remittances should be made payable to ISPO.

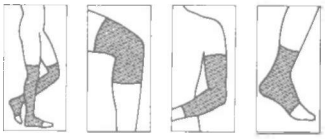
Editorial correspondence, advertisement bookings and enquiries should be directed to Prosthetics and Orthotics International, National Centre for Training and Education in Prosthetics and Orthotics, University of Strathclyde, Curran Building, 131 St. James' Road, Glasgow G4 0LS, Scotland (Tel. 041-552 4049).

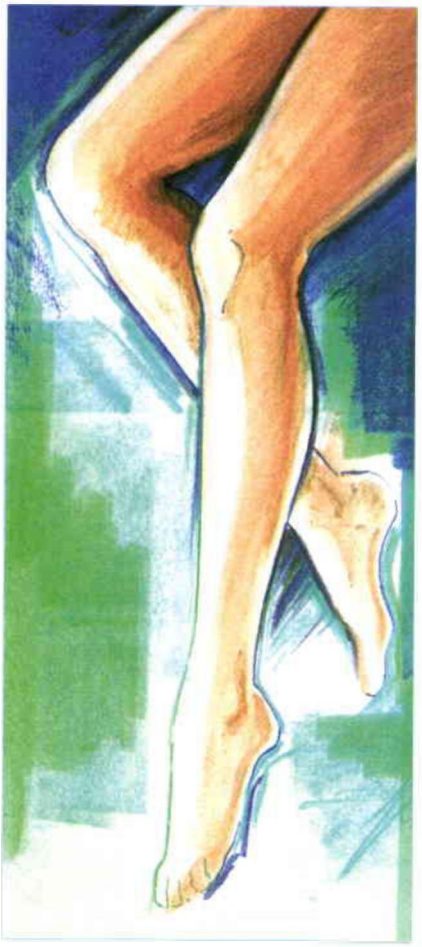
ISSN 0309–3646

Produced by the National Centre for Training and Education in Prosthetics and Orthotics, University of Strathclyde, Glasgow

Printed by David J. Clark Limited, Glasgow

Juzo® Compression Therapy for Natural Mobility





Juzo products blaze the trail – proven quality with a new look

Growing health consciousness creates new needs and new opportunities: target groups are becoming younger and more differentiated. Now there is an unmistakable trend to natural treatment, like compression therapy.

Juzo has recognized the signs of the times and is now providing modern compression garments with proven Juzo quality. At the same time, the attractive new packaging will help our retail medical supply partners to play a competitive role in this changing market.

Juzo is moreover a reliable and dynamic partner for medical supply retailers. The regular exchange of information, sales counselling and marketing support and above all the high standards of Juzo products, the result of decades of experience – all play an important role in building a relationship of cooperation and mutual trust.

Quality is a tradition at Juzo – innovations guarantee our future together.

Juzo®

Julius Zorn GmbH, 8890 Aichach/Germany
Julius Zorn Inc., Cuyahoga Falls, Ohio 44 223/USA

ISPO

Elected Members of Executive Board

W. H. Eisma (President)	Netherlands
M. Stills (President Elect)	USA
S. Heim (Vice President)	Germany
A. Jernberger (Vice President)	Sweden
V. Angliss	Australia
P. Christiansen	Denmark
T. Keokarn	Thailand
J. Vaucher	Switzerland
J. Hughes (Immediate Past President)	UK
E. Lyquist (Past President)	Denmark
G. Murdoch (Past President)	UK
J. Steen Jensen (Hon. Treasurer)	Denmark
N. A. Jacobs (Hon. Secretary)	UK

Standing Committee Chairmen and Task Officers

B. Ebskov (Finance)	Denmark
J. Hughes (Protocol and Nominations)	UK
M. Stills (Congress and Membership)	USA
J. Edelstein (International Newsletter and Journal Promotion)	USA
H. C. Thyregod (Professional Register)	Denmark
H. J. B. Day (Limb Deficient Child)	UK
V. Angliss (Publications)	Australia
B. Klasson (Socket Design)	Sweden
S. Heim/J. Hughes/G. Murdoch (Education)	Germany/UK/UK

Consultants to Executive Board

H. C. Chadderton (Consumer)	Canada
R. Henze (IVO)	Germany
J. Van Rolleghem (INTERBOR)	Belgium
J. N. Wilson (WOC)	UK

International Consultants to Executive Board

P. Kapuma	Africa
J. Craig	Central America
C. Marineck	Central and Eastern Europe
S. Sawamura	Asia
M. A. A. El-Banna	Middle East

Chairmen of National Member Societies

Australia	A. van der Borch
Austria	W. Ott
Belgium	M. Stehman
Canada	G. Martel
China	Zhongzhe Wu
Denmark	P. Christiansen
Finland	L. Nummelin
Germany	R. Baumgartner
Hong Kong	K. Y. Lee
India	M. K. Goel
Israel	M. Azaria
Japan	S. Sawamura
Korea	Vacant
Netherlands	J. C. L. van der Meer
New Zealand	W. Beasley
Norway	G. Veres
Pakistan	N. M. Akhtar
Sweden	A. Jernberger
Switzerland	J. Vaucher
UK	J. C. Peacock
USA	M. Schuch

Past Presidents

K. Jansen (1974–1977)	Denmark
G. Murdoch (1977–1980)	UK
A. Staros (1980–1982)	USA
E. Lyquist (1982–1983)	Denmark
E. G. Marquardt (1983–1986)	Germany
J. Hughes (1986-1989)	UK

Secretary

Aase Larsson	Denmark

The Journal of the International Society for Prosthetics and Orthotics

August 1991, Vol. 15, No. 2

Contents

Editorial 65

Dedication 66

ISO/ISPO classification of congenital limb deficiency 67
H. J. B. Day

Heredity and dysmorphic syndromes in congenital limb deficiencies 70
D. G. R. Evans, Y. Thakker and D. Donnai

The management of the limb deficient child and its family 78
Y. Setoguchi

The prosthetic treatment of upper limb deficiency 82
B. Curran and R. Hambrey

Structured training of children fitted with myoelectric prostheses 88
L. M. Hermansson

The use of electric elbows in the rehabilitation of children with upper limb deficiencies 93
W. F. Sauter

The stump capping procedure to prevent or treat terminal osseous overgrowth 96
J. Pfeil, E. Marquardt, T. Holtz, F. U. Niethard, E. Schneider and C. Carstens

The treatment of longitudinal radial deficiency 100
D. W. Lamb

The treatment of longitudinal ulnar deficiency 104
R. J. Lovett

The surgical treatment of congenital hand deficiency 106
P. J. Sykes

The prosthetic treatment of lower limb deficiency
R. R. Hirons, K. B. Williams, R. F. Amor and H. J. B. Day

The classification and treatment of proximal femoral deficiencies
I. P. Torode and R. Gillespie

The surgical treatment of partial tibial deficiency and ankle diastasis
W. Blauth and P. Hippe

Fibular deficiency and indications for Syme's amputation
W. L. Oppenheim

Reconstructive surgery for fibular deficiency
O. E. Shatilov, A. V. Rozkov and T. V. Cheminova

The care of the limb deficient child in Australia
L. E. Jones and A. Lipson

The care of the limb deficient child in India
V. P. Sharma

The care of the limb deficient child in Japan
S. Kakurai and M. Kida

The care of the limb deficient child in North Africa
N. Sliman, A. Mrabet, S. Daghfous and M. Douik

The care of the limb deficient child in Venezuela
F. Fernandez-Palazzi, D. P. De Gutierrez and R. Paladino

Note on situation in Argentina
G. M. Arendar and S. P. Aichenbaum

The function and operation of a parent support organization
J. Bruce

A personal experience
D. Bailey

Book Review

Proposed amendment to the Constitution

International Newsletter

Calendar of Events

Editorial

The first ISPO symposium on the Limb Deficient Child was organised by Professor Ernst Marquardt and held, appropriately, at Heidelberg in August 1988. The programme which was comprehensive, included both invited and submitted papers, and dealt with all aspects of the care of such children. It had been the intention that the proceedings would be published, but in the event the enormous content of the meeting rendered such a book impracticable, and so a special issue of this Journal was planned.

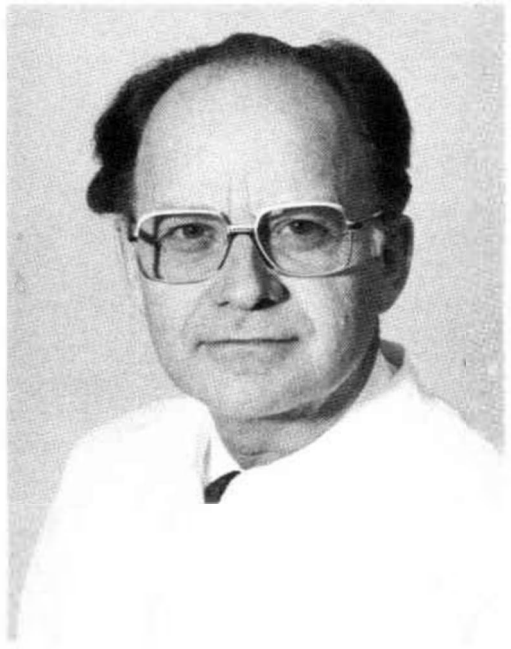
In order to provide a succinct account of the best present practice, contributions were invited from a number of experts being given tight constraints in order to avoid repetition. I take this opportunity to express my gratitude to all the authors for the way in which they accepted these strictures.

The International Standard "Method of describing limb deficiencies present at birth" (ISO 8548–1: 1989), which originated in the ISPO "Kay" committee of 1973, is used throughout and it is the intention that this will be the Journal's policy in the future.

The general articles on Classification, Dysmorphology, and the Childrens' Clinic are followed by accounts of the treatment of the individual deficiencies. However in some cases the "right" treatment in one country or continent is inappropriate elsewhere because of ethical, cultural, logistical or financial consideration. For this reason some articles are included which describe the practice in various parts of the world.

H. J. B. Day
Task Officer for The Limb Deficient Child

Dedication



Professor Dr-med Ernst Marquardt

The articles in this volume are based on some of the papers which were presented at Heidelberg in the summer of 1988. The conference was widely billed as a meeting of the International Society for Prosthetics and Orthotics but all those of us who were there knew why it was being held and what it really was — a meeting of the friends of Ernst Marquardt, held in his honour. It was a recognition of his unique personal contribution to Prosthetics and Orthotics, his teaching and of the principles of the care and management of limb deficient individuals, particularly children, which he has developed and pioneered throughout the world.

We all have our own recollections of Ernst. Amongst the many I have, perhaps the clearest is still of my first meeting with him on the morning of the 29th April 1963 when I went with my colleagues to see the work he was doing with the Dysmeliekindern at the Klinic at Heidelberg-Schlierbach. His enthusiasm and his hope for the new prostheses which he was then developing was very infectious, and I came away convinced that he had set the direction for the future development of upper limb prosthetics for all of us. More than that, at the end of five days I came away deeply impressed with the way he tackled his problems; his insistence on the innovative approach and the prosthetic developments never dominated his first and principal concern for the good of the patient as an individual, and not just the short term good either. All of us working in this field have known of cases and places where sadly such care has been overshadowed by the excitement of the research.

Over the quarter of a century that has passed while we have all grown older and some have lost the spring of youth, I can detect no change in Ernst's enthusiasm and his excitement in the pursuit of his ideals, nor in his dedicated concern for his patients; the impact of his labours has been felt all over the world. *Am Werke erkennt man den Meister.*

We all join together in wishing Ernst, his wife Weibke and their family very great happiness.

David Simpson
Edinburgh

Prosthetics and Orthotics International, 1991, 15, 67–69

The ISO/ISPO classification of congenital limb deficiency

H. J. B. DAY

Formerly of Disablement Services Centre, Withington Hospital, Manchester, UK

Abstract

This classification originally produced by the ISPO "Kay" Committee in 1973, has now with minor modifications, become an International Standard (ISO 8548–1: 1989). It is limited to those deficiencies which are failures of formation and describes them on anatomical and radiological bases only. All are divided into transverse and longitudinal, and use simple terms and descriptors.

Introduction

A logical system of classification and nomenclature is needed to facilitate scientific communication about congenital limb deficiency. The lack of a suitable system has allowed the use of the term "congenital amputation" — implying that a limb segment has been lost before birth — to be used for cases which are patently failures of formation. Furthermore any classification should use simple words capable of translation into all languages. The use of terms derived from Greek or Latin roots may sound impressively scientific, but they are both inaccurate and ambiguous, and are often misused, none more frequently than "phocomelia" which is used to describe every level and type of deficiency.

The history of classifications devised since that of Frantz and O'Rahilly (1961), including those of Burtch (1966), Henkel and Willert (1969) and the work of the ISPO "Kay" committee has been described previously by Kay (1974), Swanson (1976) and Day (1988).

The ISPO system provided the framework which enabled the Working Group of ISO Technical Committee 168 (Prosthetics and Orthotics) to set out a proposal for an International Standard. This has been accepted by the participating nations and has been published as an International Standard, ISO 8548–1: 1989 "Method of describing limb deficiencies at birth".

All correspondence to be addressed to Tithe House, Over Peover, Knutsford, Cheshire WA16 8UG, UK.

ISO 8548–1: 1989

The Standard has three constraints:–

1. The classification is restricted to skeletal deficiencies and therefore the majority of such cases are due to a failure of formation of parts.

2. The deficiencies are described on anatomical and radiological bases only. No attempt is made to classify in terms of embryology, aetiology or epidemiology.

3. Classically derived terms such as hemimelia, peromelia, etc., are avoided because of their lack of precision and the difficulty of translation into languages which are not related to Greek.

Deficiencies are described as *Transverse* and *Longitudinal.*

The former resemble an amputation residual limb, in which the limb has developed normally to a particular level beyond which no skeletal elements are present. All other cases are classed as longitudinal in which there is reduction or absence of an element or elements within the long axis of the limb.

Method of description

Transverse

The limb has developed normally to a particular level beyond which no skeletal elements exist, although there may be digital buds. Such deficiencies are described by naming the segment at which the limb terminates and then describing the level within the segment beyond which no skeletal elements exist (Table 1).

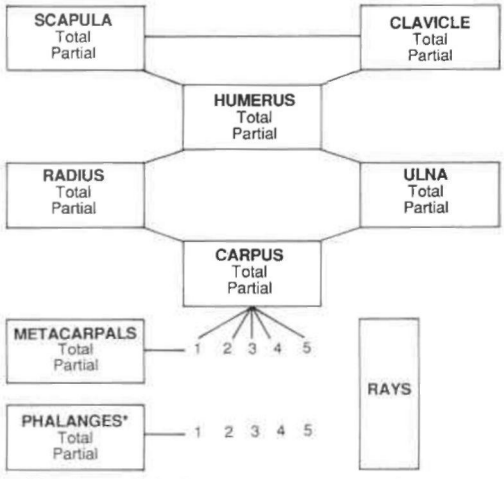
It is possible to use another descriptor in the phalangeal case to indicate a precise level of loss within the fingers.

Table 1. Designation of levels of transverse deficiencies of upper and lower limbs.

Upper limb		Lower limb
Shoulder	Total	Pelvis
Upper arm	Total Upper third Middle third Lower third	Thigh
Forearm	Total Upper third Middle third Lower third	Leg
Carpal*	Total Partial	Tarsal*
Metacarpal*	Total Partial	Metatarsal*
Phalangeal* (Finger or thumb)	Total Partial	Phalangeal* (Toe)

Notes:
(1) The skeletal elements marked * are used as adjectives in describing transverse deficiencies, e.g. transverse carpal total deficiency.
(2) Total absence of the shoulder or hemipelvis (and all distal elements) is a transverse deficiency. If only a portion of the shoulder or hemipelvis is absent, the deficiency is of the longitudinal type.

Table 2. Description of longitudinal deficiencies of the upper limb.



* The digits of the hand are sometimes referred to by name:- 1, thumb; 2 index; 3, middle; 4, ring; and 5, little (or small). For the purpose of this classification such naming is deprecated because it is not equally applicable to the foot.

Longitudinal
There is a reduction or absence of an element or elements within the long axis of the limb and in this case there may be normal skeletal elements distal to the affected bone or bones. To describe such a deficiency refer to Tables 2 and 3 and follow the procedure below:

1. Name the bones affected, in a proximo-distal sequence, using the name as a noun. Any bone not named is present and of normal form.

2. State whether each affected bone is totally or partially absent.

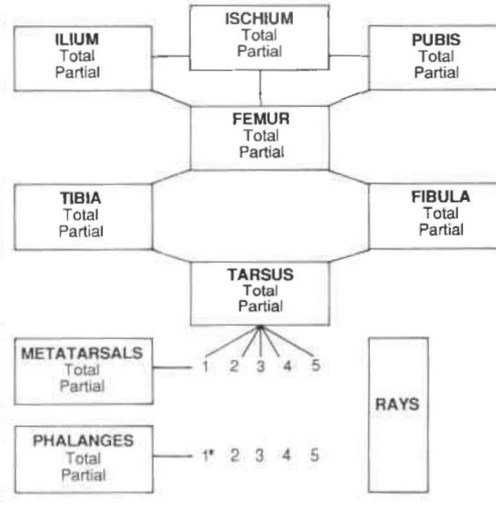
3. In the case of partial deficiencies the approximate fraction and the position of the absent part may be stated.

4. The number of the digit should be stated in relation to a metacarpal, a metatarsal and the phalanges, the numbering starting from the preaxial, radial or tibial side.

5. The term "Ray" may be used to refer to a metacarpal or metatarsal and its corresponding phalanges.

Examples of transverse and longitudinal deficiencies are shown in Figures 1 and 2, but it must be understood that the stylised representation of the limb which is used in these figures is neither part of the original ISPO "Kay" committee work nor of the new

Table 3. Description of longitudinal deficiencies of the lower limb.



* Great toe or hallux.

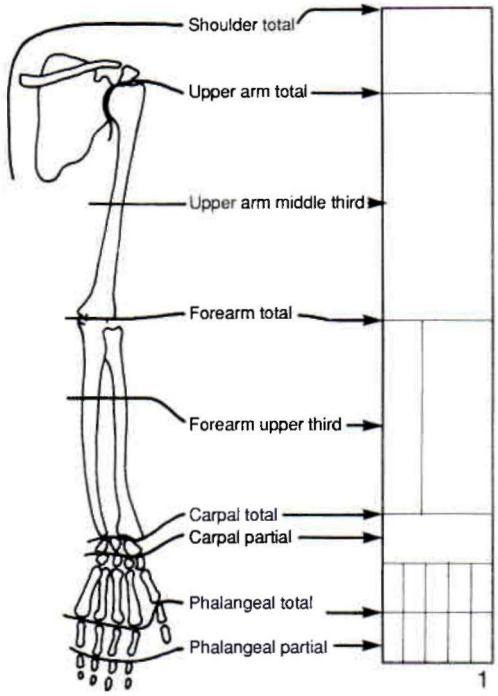


Fig. 1. Examples of transverse deficiencies at various levels, shown on the skeleton and as the author's stylised representation.

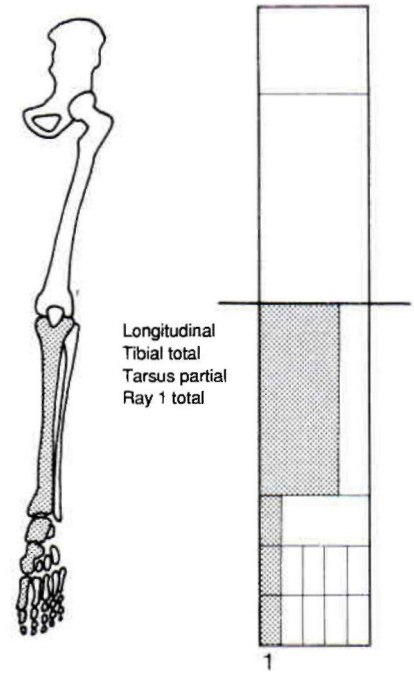


Fig. 2. Example of a longitudinal deficiency shown on the skeleton and as the author's stylised representation, showing not only the original deficiency but also the treatment by knee disarticulation.

International Standard, but the author has found it to be the most useful way of illustrating deficiencies in clinical notes and it can be used to indicate some treatment as well as the deficiency.

Acknowledgement

Parts of ISO 8548–1: 1989 are reproduced here with the permission of the International Organisation for Standardisation (ISO). Copies of this standard are available from the ISO Central Secretariat, Case Postale 56, CH-1211 Geneva 20, Switzerland or from any ISO member body.

REFERENCES

BURTCH, R. L. (1966). Nomenclature for congenital skeletal limb dificiencies, a revision of the Frantz and O'Rahilly classification. *Artificial Limbs,* **10(1)**, 24–35.

FRANTZ, C. H., O'RAHILLY, R. (1961). Congenital skeletal limb deficiencies. *J. Bone Joint Surg.*, **43A**, 1202-1204.

DAY, H. J. B. (1988). Nomenclature and classification in congenital limb deficiency. In: Amputation surgery and lower limb prosthetics. Edited by Murdoch. G. Blackwell. — Edinburgh, Blackwell Scientific. p271-278.

HENKEL, H. L., WILLERT H. G. (1969). Dysmelia, a classification and a pattern of malformation of congenital limb deficiencies. *J. Bone Joint Surg.*, **51B**, 399–414.

ISO 8548–1: 1989. Prosthetics and Orthotics — Limb deficiencies — Part 1: Method of describing limb deficiencies present at birth.

KAY, H. W. (1974). A proposed international terminology for the classification of congenital limb deficiencies. *ICIB,* **13(7)**, 1–16.

KAY, H. W. (1975). The proposed international terminology for the classification of congenital limb deficiencies. The recommendations of a working group of ISPO. *Dev. Med. Child Neurol. Suppl.*, **34**, 1–12.

SWANSON A. B. (1964). A classification for congenital malformations of the hand. *N. J. Bull., Acad. Med.*, **10**, 166–169.

SWANSON A. B. (1976). A classification for congenital limb malformations. *J. Hand Surg.*, **1**, 8–22.

Prosthetics and Orthotics International, 1991, 15, 70–77

Heredity and dysmorphic syndromes in congenital limb deficiencies

D. G. R. EVANS, Y. THAKKER and D. DONNAI

Department of Medical Genetics, St. Mary's Hospital, Manchester, UK

Abstract

Isolated limb deficiencies are usually sporadic occurrences. However, if they are associated with other abnormalities or a family history, the risk to future pregnancies may be as high as 50%. A thorough history, examination and investigation of the baby as well as the parents is essential before assessing this recurrence risk. The syndromes associated with limb deficiencies are presented.

Introduction

Limb deficiency has an incidence of approximately 1:2,000 births and in some families there may be a significant recurrence risk (Calzorali *et al.*, 1990). After a parent has recovered from the initial shock of finding their baby has a problem, two important questions arise. The first is about the implications of the anomaly for the child. The second question involves the risks of the problem happening again.

To answer this, it is necessary to determine whether the baby's anomalies fit into a "pattern" which constitute a syndrome. A pregnancy history, including drug or alcohol exposure, should be taken, as should a full family history concentrating on limb abnormalities. The baby should then be examined for dysmorphic features arising from abnormal embryogenesis. Recognition of a pattern of malformations may enable conclusions to be drawn about the mechanism and timing of the anomalies. Certain investigations such as blood count, bone marrow aspirate and chromosome analysis may be needed in addition to X-rays. Finally the parents should be examined and if necessary X-rayed for signs of minor anomalies which may represent reduced expression of a dominant gene.

The classification of limb deficiencies has previously been made on anatomical grounds (O'Rahilly, 1969). To illustrate this article we have made a causal and genetic classification (Tables 1 and 2) but this is not meant to replace the former, as the majority of defects are sporadic with ill-explained mechanisms.

Anomalies such as duplication, polydactyly, brachydactyly, syndactyly and the pterygium syndromes are not discussed, neither are skeletal dysplasias. We have concentrated on conditions presenting as limb reduction or deficiency.

Single gene disorders

Disorders caused by a defect in a single gene follow the patterns of inheritance described by Mendel. There may be heterogeneity within a particular diagnostic category. For example the typical split hand anomaly can be inherited in any one of the three Mendelian ways: autosomal dominant, autosomal recessive or X-linked recessive, or it may be sporadic or part of a syndrome. The family pedigree and clinical presentation may allow confident counselling.

Autosomal dominant disorders

Autosomal dominant disorders affect both males and females and can often be traced through many generations of a family. Affected people are heterozygous for the abnormal allele and transmit the gene for the disorder on average to half their offspring, whether male or female. Estimation of risk is apparently simple but in practice, factors such as variable

All correspondence to be addressed to Dr. D. G. R. Evans, Senior Registrar, Department of Medical Genetics, St. Mary's Hospital, Hathersage Road, Manchester M13 0JH, UK.

Table 1: Syndromes presenting with limb deficiency.

Aetiology	Syndrome	Limb deficiency
Single gene disorders:		
Autosomal dominant:	EEC	split hand/foot (ectrodactyly)
	Adams Oliver	transverse defect
	Holt Oram	longitudinal all degrees
	LADD	absence thumb/radius
	Duane	absence to hypoplasia radius/thumb
	Split hand/foot	split hand/foot
Autosomal recessive:	Roberts	longitudinal affecting whole limb
	Grebe	hypoplastic radii, ulnae, tibiae
	Fanconi	hypoplasia to absence thumb/radius
	TAR	absence of radius — thumb present
	AASG	hypoplasia of radius
	Miller	hypoplasia to absence ulna/5th ray
	Weyers	oligodactyly, hypoplasia ulnar ray
	Split hand/foot	split hand/foot
X-linked recessive:	Split hand/foot	split hand/foot
Chromosomal:	trisomy 18	longitudinal all degrees
	trisomy 13	radial absence
	13q−	hypoplastic thumb
	4q−	hypoplasia/absence thumb
Drugs:	alcohol	longitudinal all degrees
	thalidomide	longitudinal all degrees
	aminopterin	hypoplasia (forearm), hypodactyly
Infection:	varicella	hypoplasia and hypodactyly
Aberrant development:	MZ twinning	transverse defect
	amniotic bands	transverse defect
Sporadic syndromes: (unknown cause)	aglossia-adactyly	adactyly
	Poland/Moebius	hypoplasia forearm, oligodactyly
	spleno-gonadal	transverse defect
	VATER	absent radius
	Cornelia de Lange	micromelia, hypoplasia ulna
	FFU (also A.R.)	hypoplasia femur, fibula, ulna
Associated with maternal diabetes:	Caudal dysplasia	hypoplasia lower limb
	femoral hypoplasia + unusual facies	hypoplasia/absence femur, fibula and humerus

expression and penetrance may cause difficulties in counselling. The severity of many dominant conditions vary considerably among affected members within a family. The likely severity is difficult to predict and a mildly affected parent may have a severely affected child.

Adams Oliver syndrome: A parent with minor terminal reductions of toes and fingers may have a child with severe limb defects such as bilateral transverse leg deficiencies (Fryns, 1987). The associated scalp defects are midline skin defects located on the vertex with occasional defects of the skull and meninges.

Holt-Oram syndrome: There is a wide variability of expression of both the cardiac and upper limb anomalies (Fig. 1). There are all gradations of defects in the upper limb ranging from clinodactyly and carpal fusion to more severe longitudinal deficiencies, the most typical being a thumb anomaly. The cardiac lesion, may be an atrial septal defect (common), ventricular septal defect, patent ductus arteriosus or mitral-valve prolapse. There is no correlation between the severity of

Fig. 1. Holt Oram syndrome: note bilateral radial absence.

the limb defect and the cardiac defect. Before a new mutation is presumed to have occurred and a low recurrence risk is given to apparently normal parents of an affected child, detailed physical examination with X-rays of wrist, hand and arms and cardiac assessment with echocardiography are necessary.

EEC syndrome: Each of the three main features of ectrodactyly, ectodermal dysplasia and clefting syndrome shows a variable degree of expressivity. The ectodermal part of this syndrome involves the hair (dry and sparse with absent eyebrows and eyelashes), teeth (small or absent) and nails (brittle and ridged). Limb abnormality consists of defects in midportion of hand and feet, varying from syndactly to a split hand/foot anomaly. Mental development is usually normal and facial clefting, is common but not an essential part of the syndrome.

LADD (lacrimo-auriculo dento-digital) syndrome: Phenotypic variation is seen. (Thompson *et al.*, 1985). The characteristic features of this condition are absence of lacrimal puncta/canaliculi leading to epiphora and chronic eye infections, cup-shaped or malformed ears with sensoneural or conductive deafness, abnormalities of the teeth and radial ray defects of variable severity.

Table 2. Classification of syndromes by anatomical criteria.

Limb deficiency		*Syndromes*
a) Transverse defect: previously known as:		MZ twinning
	amelia	Adams Oliver
	hemimelia	amniotic bands
	acheiria	aglossia adactyly
	adactylia	EEC
	ectrodactyly	varicella
	apodia	alcohol
b) Proximal longitudinal: absent radius, thumb present hand attached at shoulder foot attached to hip		thalidomide Roberts Grebe aminopterin TAR varicella
c) Longitudinal	i) radial ray:	VATER Holt Oram Nager Fanconi Levy Hollister trisomy 18 Duane AASG
	ii) ulnar ray:	Cornelia de Lange Miller FFU Weyers
d) Split hand/foot:		Cornelia de Lange EEC split hand/foot XLR, AD and AR

Duane syndrome: Radial ray defects associated with Duane anomaly follow an autosomal dominant pattern in some families (MacDermot and Winter, 1987). The radial defects range from thenar eminence hypoplasia to radial absence. The Duane anomaly is an unusual congenital form of strabismus characterised by limitation of abduction in association with retraction of the globe and narrowing of the palpebral fissures on adduction.

Split hand/foot: Autosomal dominant inheritance is the commonest mode of transmission in familial isolated split hand/split foot anomaly. The anomaly may be of the lobster claw variety (absence of central rays) or monodactyly type (deficiency of radial rays with no cleft). Gradations between these types occur and cases of each type are seen in some families with the appearance of skipped generations.

Autosomal recessive disorders

Autosomal recessive disorders occur in a person whose healthy parents carry the same recessive gene. The risk of recurrence for future offspring of such parents is 1 in 4 (25%). Consanguinity increases the risk of a recessive disorder because both parents are more likely to carry the defective gene which has been inherited from a common ancestor.

The following are examples of some of the conditions following this pattern of inheritance.

Roberts syndrome: The main clinical features are severe symmetrical shortening of the limbs with end longitudinal deficiency (Fig. 2), a characteristic face with hypertelorism, severe cleft lip, prominent premaxilla, mid-face capillary haemangioma, cloudy corneas or cataracts and dysplastic ears. Many affected infants die in the newborn period, survivors are mentally retarded. Clinical and genetic data suggest it is probably the same condition as has been described as SC phocomelia. Chromosome analysis in both conditions shows premature centromeric separation (Romke *et al.*, 1987).

Grebe syndrome: This condition is characterised by severe shortening of the upper

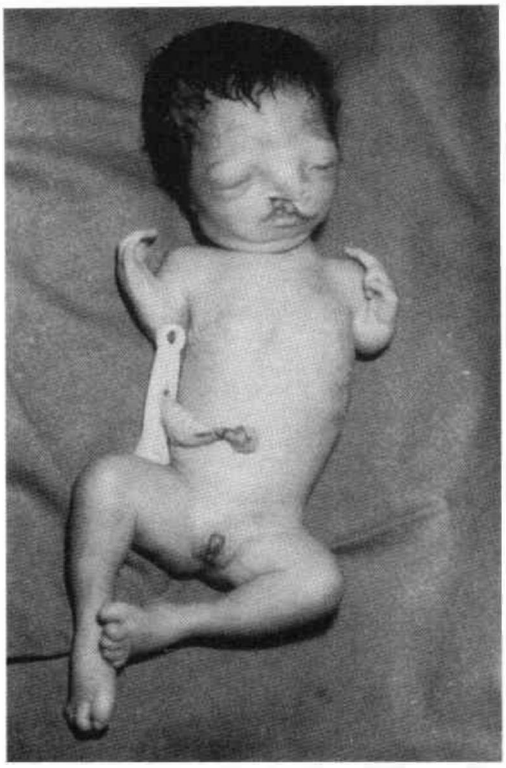


Fig. 2. Roberts syndrome: note characteristic facial pattern with cleft palate and symmetrical deficiency worse in upper limbs.

and lower limbs so that the eventual height can be as small as 3 feet. The hand and fingers are particularly tiny, the digits resembling stubby toes. The head, trunk, and intelligence of survivors are normal.

Fanconi anaemia: This disorder presents as a pancytopenia (mean age of onset — 8 years), associated with a variety of congenital malformations. The malformations affect one or more systems, including the skin, skeletal, ocular, auditory, renal, genital and central nervous systems. Radial defects comprising hypoplasia or absence of the thumb, first metacarpal and radius are the most common skeletal malformations (Glanz and Fraser, 1982). This variation of number and severity of the congenital malformations precludes the establishment of hard and fast diagnostic criteria on clinical grounds only. In recent years, cytogenetic studies have demonstrated that affected persons have an increased frequency of chromosomal breaks, even in the pre-anaemic stage. The chromosomal breaks can also be demonstrated in the cultured amniotic fluid cells allowing prenatal diagnosis of infants at risk (Auerbach *et al.*, 1985).

TAR (thrombocytopenia and absent radius): The clinical features include haematological abnormalities (mainly of platelets), skeletal abnormalities (primarily of both arms and legs), cardiac abnormalities (particularly tetralogy of fallot and atrial septal defect), and cow's milk allergy. The most striking skeletal abnormality is bilateral radial absence with preservation of the thumbs. The hypomegalokaryocytic thrombocytopenia is of early onset. Anaemia, eosinophilia and leukaemoid granulations may be seen. If the infant survives, the haematological features become less severe.

AASE — triphalangeal thumb with congenital anaemia: A congenital erythroid hypoplastic anaemia is associated with triphalangeal thumbs which lie in the same plane as the other digits. Sloping narrow shoulders, hypoplasia of the radius and radio-ulnar synostosis have been noted in some cases. The anaemia usually responds to steroids and improves with age. There is controversy about the existence of this condition as a separate entity from Blackfan Diamond congenital erythroid hypoplastic anaemia (Alter, 1978).

Weyers oligodactyly: The main features of this syndrome which was first described by Weyer are deficiency of the ulnar and fibular rays, antecubital pterygia, reduced sternal segments and malformations of the kidney and spleen with cleft lip and palate. Two affected siblings were reported by Elejalde *et al.*, (1985) suggesting autosomal recessive inheritance.

Miller (postaxial acrofacial dysostosis) syndrome: The characteristic features are postaxial limb deficiency, cup-shaped ears and malar hypoplasia. The limb defects are distinctive, consisting of an absence or incomplete development of the fifth digital ray of all four limbs (Donnai *et al.*, 1987). Most cases have shortened forearm with ulnar hypoplasia. The facial features are similar to Nager syndrome in which the limb deficiencies are preaxial.

Split hand/split foot anomaly: Recessive inheritance is less common than the dominant type but has been observed in inbred communities (Freire Maia, 1971). It should be diagnosed with caution because of variable expression of the autosomal dominant type.

X-linked recessive disorders

In X-linked recessive disorders the mutant gene is on the X chromosome, hemizygous males (with the mutant gene on their single X chromosome) are affected, heterozygous females (=Carriers) are usually healthy but may occasionally show some features of the condition. A female carrier will transmit the disorder on average to half her sons, and half her daughters will be carriers. All the daughters of an affected male are obligate carriers and none of the sons are affected.

Split hand/split foot anomaly: Ahmed *et al.*, (1987) reported a large (7 generations) inbred kindred from Pakistan in which an isolated type of split hand/split foot anomaly is transmitted as an X-linked recessive disorder. Affected males show full expression of the trait, carrier females show only features such as syndactyly. There is no male to male transmission.

Chromosomal abnormalities

The correct amount of chromosomal material is essential for the normal formation and function of a baby, any imbalance usually results in multiple congenital malformations and mental retardation. Common groups of chromosomal abnormalities include trisomies (extra chromosome present), deletion and duplication (part of chromosome is missing or duplicated) and mosaics (abnormal chromosomal pattern present in only some cells).

There are a few well recognised syndromes associated with autosomal trisomies — Down's syndrome (Trisomy 21), Edward's syndrome (Trisomy 18) and Patau's syndrome (Trisomy 13). Congenital limb deficiencies have been described in Edward's and Patau's syndrome. At least 10% of cases of Edward's have radial and or thumb absence, the characteristic hand deformity being tight flexion of the fingers with the 2nd and 5th digits overlapping the 3rd and 4th. In Patau's syndrome the characteristic hand deformity is post-axial polydactyly, absence of the radial rays although the radius is sometimes present. The majority of trisomies arise from non-disjunction at meiosis, and the recurrence risk for these is low.

Sometimes only part of a chromosome is missing or duplicated. There are many such conditions and only a few are common enough to have a recognisable "named" pattern of malformations. Chromosomal analysis should therefore be considered in any baby with multiple congenital abnormalities, particularly if associated with a low birth weight. Limb deficiencies with hypoplasia to absence of the thumb have been described in cases with partial deletion of the long arm (q) of chromosomes number 13 and 4. Duplication of part of chromosome 3 gives an appearance similar to de Lange syndrome.

As with the translocation trisomies, chromosomal structural abnormalities may arise as *de novo* events or from a balanced rearrangement in one of the parents. Parental karyotypes are indicated for any child with a chromosomal abnormality other than the regular trisomies. Extended family studies will be required if a parent has a balanced structural abnormality. The recurrence risk varies with the type of chromosomal rearrangement and sex of the carrier. Prenatal diagnosis with amniocentesis or chorionic villous biopsy should be offered to all women whose pregnancies are at increased risk.

Drugs

Skeletal limb deficiencies have been produced in the rat and mouse experimentally using a number of drugs in common use today (Freire Maia, 1969). There are only a few agents which have been firmly implicated in man. The best known is thalidomide (Millen, 1962), which may cause a wide range of longitudinal defects up to complete absence of all four limbs. Administration between the 38th and 47th day of pregnancy is most dangerous. Thalidomide still has an important role in leprosy therapy and we may not have seen the last affected infant with thalidomide embryopathy.

The folate antagonists aminopterin (Thiersch, 1952) and methotrexate (Milunsky *et al.*, 1968) have been associated with limb deficiency. Aminopterin was used as an abortifacient drug and anomalies noted

included growth deficiency, cranial dysplasia with broad nasal bridge and short limbs especially in the forearm, with hypodactyly.

The foetal alcohol syndrome occurs when the foetus is subjected to large quantities of alcohol in pregnancy. Infants are usually small hypotonic and jittery. They are microcephalic with short palpebral fissures and a smooth under developed philtrum. There may be cardiac lesions. Limb deficiency is rare but can be severe with reduction or even total absence (Pauli and Feldman, 1986).

Infection

The only infection definitely associated with limb defects is varicella. Infection in early pregnancy can lead to a number of anomalies including: cicatricial skin, chorioretinitis, growth and mental deficiency and seizures. The limb may be hypoplastic with rudimentary digits, perhaps because of viral or inflammatory damage to the nerves of the developing limb. Only a small percentage of mothers with proven infection have an affected baby (Paryani and Arvin, 1986).

Other known mechanisms

Monozygous twinning: This may cause a number of effects by various means (Schinzel *et al.*, 1979). Death of one twin may lead to thromboplastin or embolic release into the circulation of the co-twin causing a transverse defect by vascular disruption.

Amniotic bands: These have long been implicated in the production of limb deficiencies and are often called Streeter's bands (Streeter, 1930). The exact mechanism of their origin is still unclear. Limb defects include constriction rings, secondary syndactylies and amputations.

Poland and Moebius anomalies: Poland anomaly consists of unilateral absence of the pectoralis muscle and ipsilateral symbrachydactyly. Transverse defects especially distally may co-exist. In the Moebius anomaly there is unilateral or bilateral palsy of the sixth and seventh cranial nerves. The third, fifth, ninth and twelfth may also be affected. Limb deficiencies are similar to those that occur in Poland anomaly. There can be considerable overlap between the two conditions.

A common mechanism for the two has recently been proposed (Bouwes Bavinck and Weaver, 1986). This involves disruption of the blood flow at a critical stage of embryogenesis (day 37–42). If this occurs in the subclavian artery it will result in Poland anomaly and if it occurs in the basilar or vertebral arteries Moebius anomaly results. If interruption occurs at both sites the conditions could co-exist, as they often do clinically.

Other syndromes

VATER: The non-random association of vertebral defects, anal atresia, tracheo-oesophageal fistula, radial aplasia, and renal anomalies, has been reported (Temtamy and Miller, 1974). This has been extended to *VACTERL* more recently to incorporate cardiac and other limb abnormalities. These include hypoplasia of the humerus andy varying degrees of aplasia in the lower limb (Fig. 3) (Fernbach and Glass, 1988). This disorder is sporadic and of unknown cause.

Cornelia de Lange: This is a very well characterised condition. Infants are small, and mentally retarded with microbrachycephaly. They have bushy eyebrows and synophrys and a small nose with anteverted nostrils. Micromelia affects the legs predominantly, however there can be severe longitudinal deficiency and oligodactyly in the upper limbs and the ulnar ray is often involved. The condition is sporadic but because it occurs relatively frequently and has all the hallmarks of a chromosome deletion syndrome, it may well be caused by gene imprinting (Hall, 1990). Duplication of the chromosome 3q25–29 band gives a very similar phenotype (Wilson *et al.*, 1978).

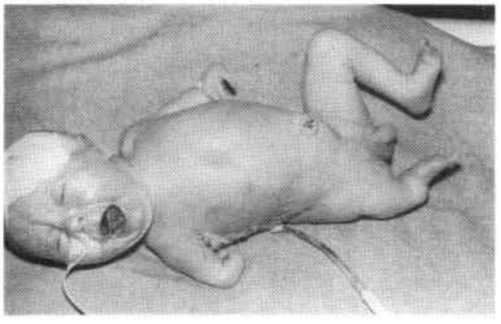


Fig. 3. VATER: note assymmetrical limb deficiency

Counselling

The majority of limb defects, especially those affecting one limb only, are sporadic and parents can be given a low recurrence risk. However there are pitfalls to this as the occurrence of isolated limb deficiencies may be due to autosomal recessive genes (Hecht, 1981). The presence of associated abnormalities may allow a diagnosis of a recognised syndrome. There are varying reports as to the frequency of these, Calzorali *et al.* (1990) estimate 12% with half representing known syndromes. Generally parents can be given a high (50–25%), medium (10–5%), or low recurrence risk. The Mendelian conditions would give a high risk. The medium risk would be appropriate when it was thought that complex genetic mechanisms or multifactorial inheritance was involved. Low risk is usually appropriate to isolated transverse deficiency, ulnar absence and quadruple amelias. A low risk can also be given if a known mechanism such as varicella or a drug was involved. Care must be taken however with the foetal alcohol syndrome as there are often several affected siblings with markedly different phenotypes. Another difficult area is that of a consanguineous marriage in a situation where a low risk would normally be given (Freire Maia, 1969). Caution is necessary in such circumstances.

Parents seen at or soon after the birth, by a clinical geneticist, should be seen again a few months later for formal genetic counselling. Virtually all parents experience some guilt feelings and will have their own (often far-fetched) explanations for why the abnormality has occurred. It is important to allow parents the opportunity to vent these concerns. At the formal counselling session parents may be looking to the future and wish to discuss the recurrence risks and possible prenatal diagnosis. Genetic counselling aims to give information to allow parents an informed choice and does not presume to advise people on a "correct" course of action. Ultrasound has markedly improved the possibilities of prenatal diagnosis and when appropriate, a detailed scan should be offered.

Conclusion

The authors have outlined the syndromes associated with limb deficiency. Although these represent only a small proportion of the total number of individuals with limb deficiency it is important that these are identified in order to give accurate genetic counselling to relatives.

REFERENCES

Ahmed, M., Abbas, M., Haque, S., Flatz, G. (1987). Chromosally inherited split hand/split foot anomaly in a Pakistani kindred. *Hum. Genet.*, **75,** 169–173.

Alter, B. P. (1978). Thumbs and anaemia. *Pediatrics,* **62,** 613–614.

Auerbach, A. D., Sagi, M., Alder, B. (1985). Fanconi anemia: prenatal diagnosis in 30 fetuses at risk. *Pediatrics,* **76,** 794–800.

Bouwes Bavinck, J. N., Weaver, D. D. (1986). Subclavian artery supply disruption sequence: hypothesis of a vascular etiology for Poland, Klippel-Feil, and Moebius anomalies. *Am. J. Med. Genet.*, **23,** 903–918.

Calzorali, E., Manservigi, D., Garani, G. P., Cocchi, G. Magagni, C., Milan, M. (1990). Limb reduction defects in Emilia Romagna, Italy: epidemiological and genetic study in 173,109 consecutive births. *J. Med. Genet.*, **27,** 353–357.

Donnai, D., Hughes, H. E., Winter, R. M. (1987). Postaxial acrofacial dysostosis (Miller) syndrome. *J. Med. Genet.*, **24,** 422–425.

Elejalde, B. R., de Elejalde, M. M., Booth, C. Kaye, C., Hollison, L. (1985). Prenatal diagnosis of Weyer's syndrome (deficient ulnar and fibular rays with bilateral hydronephrosis). *Am. J. Med. Genet.*, **21,** 439–444.

Fernbach, S. K., Glass, R. B. J. (1988). The expanded spectrum of limb anomalies in VATER association. *Pediatr. Radiol.*, **18,** 214–220.

Freire Maia, N. (1969). Congenital skeletal limb deficiencies: a general view. *Birth Defects OAS,* **5 (3),** 7–13.

Freire Maia, A. (1971). A recessive form of ecterodactyly and its implications in genetic counselling. *J. Hered.*, **62,** 53.

Fryns, J. P. (1987). Congenital scalp defects with distal limb anomalies. *J. Med. Genet.*, **24,** 493–496.

Glanz, A., Fraser, F. C. (1982). Spectrum of anomalies in Fanconi anaemia. *J. Med Genet.*, **19,** 412–416.

Hall, J. (1990). Genomic imprinting: review and relevance to human diseases. *Am. J. Hum. Genet.*, **46,** 857–873.

Hecht, J. T., Scott, C. I. (1981). Recurrent unilateral hand malformations in siblings. *Clin. Genet.*, **20,** 225–228.

MacDermot, K. D., Winter, R. M. (1987). Radial ray defect and Duane anomaly: report of a family with autosomal dominant transmission. *Am. J. Med. Genet.*, **27**, 313–319.

Millen, J. W. (1962). Thalidomide and limb deformities. *Lancet*, **II**, 599–600.

Milunsky, A., Graf, J. W., Gaynor, M. F. (1968). Methotrexate induced congenital malformations with a review of the literature. *J. Pediatr.*, **72**, 790–795.

O'Rahilly, R. (1969). Nomenclature and classification of limb anomalies. *Birth Defects OAS*, **5 (3)**, 14–16.

Paryani, S. G., Arvin, A. M. (1986). Intrauterine infection with varicella zoster virus after maternal varicella. *N. Engl. J. Med.*, **314**, 1542–1546.

Pauli, R. M., Feldman, P. F. (1986). Major limb malformations following intrauterine exposure to ethanol; two additional cases and literature review. *Teratology*, **33**, 273–280.

Romke, C., Froster-Iskenius, U., Heyne, K., Hohn, W., Hof, M., Grzejszcyk, G., Rouskolb, R., Rehder, H., Schwinger, E. (1987). Roberts syndrome and SC phocomelia. A single gene entity. *Clin. Genet.*, **31**, 170–177.

Schinzel, A. A. G. L., Smith, D. W. Miller, J. R. (1979). Monozygotic twinning and structural defects. *J. Pediatr.*, **95**, 921–930.

Stevenson, R. E., Jones, K. L., Phelan, M. C., Jones, M. C., Barr, M., Clericuzio, C., Herley, R. A., Benirschke, K. (1986). Vascular steal: the pathogenetic mechanism producing sirenomelia and associated defects of the viscera and soft tissues. *Pediatrics*, **78**, 451-457.

Streeter, G. L. (1930). Focal deficiencies in fetal tissue and their relation to intrauterine amputation. *Contra. Embryol. Carneg. Inst.*, **22**, 1–44.

Temtamy, S. A., Miller, D. J. (1974). Extending the scope of the VATER association: definition of the VATER syndrome. *J. Pediatr.*, **85**, 345–349.

Thiersch, J. B. (1952). Therapeutic abortions with a folic acid antagonist, 4 aminopteroyl glutamic acid (4 amino PGA) administered by the oral route. *Am. J. Obstet. Gynecol.*, **63**, 1298.

Thompson, E., Pembrey, M., Graham, J. M. (1985). Phenotypic variation in LADD syndrome. *J. Med. Genet.*, **22**, 382–385.

Wilson, G. N., Hieber, V. C., Schmickel, R. D. (1978). The association of chromosome 3 duplication with Cornelia de Lange syndrome. *J. Pediatr.*, **93**, 783–788.

Prosthetics and Orthotics International, 1991. 15, 78-81

The management of the limb deficient child and its family

Y. SETOGUCHI

Child Amputee Prosthetics Project, Shriners Hospital, Los Angeles, USA.

Abstract

A properly constituted clinic team needs to be involved as soon as possible after the birth of a child with a limb deficiency, to answer the parents' questions, counsel them and the family, and plan the treatment programme for the child so that they can properly assist the child to become a productive, self-sufficient member of society.

The management of the limb deficient child

The birth of a child with congenital anomalies is a traumatic event for parents who have been expecting a normal child. This is compounded when the physical defect is visible such as in the case of a limb deficiency. The obstetrician or paediatrician is often unprepared and the parents and other immediate family members need to be seen as soon as possible to begin the treatment programme. It is this philosophy that physicians like Ernst Marquardt and Milo B. Brooks, emphasised in their concepts of the management of the limb deficient child. They felt that it was not just the physical treatment of the child but the positive bonding of the parents to their child that ultimately led to a healthy, productive member of our society, instead of a "handicapped" person.

The team approach, the involvement of a multi-disciplinary group of professionals focusing on all aspects of the child's needs, was advocated by Marquardt, Brooks and others. The team's emphasis was on the needs of the whole child and his family. The child's medical, genetic, and sometimes surgical needs are all important in the treatment programme as are the prosthetic and therapy requirements. It is most important to co-ordinate all of the above treatment modalities with evaluation and treatment of the psychosocial needs of the patient and his/her family. The team's responsibility is to ensure that all aspects of the child are assessed and appropriate therapy provided.

No parents imagine that their baby will be born with a defect. Therefore, regardless of the severity of the problem, all parents experience shock at the birth of the limb deficient child. They may experience other emotional stresses, such as sadness, anger, embarrassment, guilt, feelings of isolation, feelings of failure as parents, denial and unhappiness. How the parents are helped to accept that these are normal reactions and to resolve these feelings can make a great difference in their response to the child's limb deficiency. The longer the parents are left to experience their feelings without appropriate resolution, the more likely they will develop unhealthy attitudes and reactions. This in turn can have very harmful effects on the parent/child bonding.

The initial evaluation should be made as soon as possible. In fact, the first consultation is often in the newborn nursery. The parents need to have their questions answered as soon as possible and to be given a chance to vent their feelings. If a visit to the hospital by the clinic team, usually physician and social worker, is not possible or necessary, then a clinic appointment is scheduled as soon as possible. As soon as a referral is made, the clinic social worker contacts the family to inform them of the functions and procedures of the clinic. Questions regarding the initial evaluation, as well as the financial aspect of the treatment programme are answered.

A booklet entitled "The Child with a Limb Deficiency: A Guide for Parents" is sent to the family. The subjects covered in this short and concise pamphlet include: the problem at birth;

All correspondence to be addressed to Dr. Y. Setoguchi, Medical Director, Child Amputee Prosthetics Project, Shriners Hospital for Crippled Children, 3160 Geneva Street, Los Angeles. CA 90020 - 1199, USA.

fears about the future; reactions of others; self-acceptance; your child's questions; school and potential concerns; adolescence; and vocational considerations. It is not intended to cover all of the subjects thoroughly but to answer many of the questions that the new parents may have. The booklet also helps the family prepare for the first appointment and raises other questions that the team will need to address. The acceptance of the "physical difference" and the family's ability to deal with the limb deficiency (ies) is critical to the child's future treatment efforts.

The first visit is a very important part of the treatment programme. The parents have many questions that need to be clarified. It is important that the team address these issues in a simple, clear manner. The first question usually is "What caused this to happen?" Often the parents are looking for someone to blame, other than themselves. They wonder about medicines they may have taken, chemicals or environment substances they may have been exposed to, and in one group, the effects of exposure of the mother to lunar or sonar eclipses. They wonder whether the child has other abnormalities. Of particular concern to the family is whether the child will be of normal intelligence.

From a functional standpoint, the parents are concerned whether the child will be able to perform self-care activities and be able to compete effectively in school and later in vocational endeavours.

The initial consultation should be done by a multidisciplinary team familiar with the needs and treatment of children with limb deficiency. The usual team consists of a paediatrician, (usually the team leader), orthopaedic surgeon, physiatrist, medical social worker, occupational therapist, physical therapist, prosthetist, geneticist, and rehabilitation engineer. Not all of these individuals are involved in the clinic assessment at each visit, but are needed to maintain the maximum and most up to date treatment programme for the limb deficient child.

The assessment should include not only the child's medical history and a thorough physical examination, but also the psychosocial status of the parents including the family's financial and emotional ability to support a treatment programme. A careful and complete explanation of the various prosthetic fittings and components available must be given. They should also be given information about how the child would function if no prosthesis is fitted. It should be emphasised that therapy is available to allow the child to become as independent as possible with and without prosthetic fitting. Some children with limb deficiencies are more functional using the residual limbs or their feet than they can be with prostheses. The acceptance of the prosthesis by the child and family depends on their understanding of its purpose and function as well as their approval of the ultimate fitting.

A complete history and physical examination of the patient is essential. Today, a number of syndromes, some with genetic or hereditary implications are recognised and these must be identified. At times a genetic consultation is important. This is especially true when multiple anomalies, are noted. Although this article is not intended to be specific, two conditions need to be mentioned. Radial deficiencies, both unilateral and bilateral and tibial absence are known to have hereditary implications and so patients with these conditions need to be referred to a geneticist. Other medical problems associated with limb deficiencies include craniofacial anomalies; musculoskeletal anomalies; cardiac and haematopoetic disorders.

Questions frequently asked about treatment include either transplantation or reconstruction of a limb. The ultimate hope of any parent is to be able to provide a "normal" arm or leg for their child. Even in a prosthetic device many parents are seeking a normal "flesh and blood" extremity. These concerns must be addressed by the team and correct and truthful answers must be provided.

The assessment of the "total" child is an absolute necessity in providing the best rehabilitation, including prosthetic treatment for the limb deficient child who is the "core" or central figure in the treatment programme. Initially treatment decisions are made by the parents, but as soon as the child is mature enough to participate in the decision making process he should be allowed to do so, and be actively involved in any decision regarding prosthetic fitting, therapy and other treatment programmes.

The parents are the first to learn about the

birth defect(s). How they cope with this initial shock will have a great deal to do with their abilities to raise their child with a healthy attitude about its self-image and esteem. The team must allow the parents to express their feelings and to develop positive approaches to manage the child's needs as he/she grows. The parents need to know that they are not the only ones with a child with limb deficiencies. Introduction to other families or to support groups is very helpful.

Concern should not be limited only to the immediate family. There are many family members who take part in the child's care and, therefore, need to be involved in the evaluation and subsequent treatment programme. Siblings are often the neglected members of the family. The patient initially requires considerable time and effort from the parents, and other siblings, especially the next older child are affected. Until the patient was born the older sibling was the "baby" of the family and had the attention of the parents. A normal newborn baby requires considerable parental attention but a child with a birth defect demands more. The older child therefore feels left out and may even feel that he or she was to blame for the birth defect. The team's attention to the older sibling and his inclusion in the treatment programme often will provide a valuable ally for the patient in later years.

Relatives too are important in the evaluation process. They may not actually participate in the decision making but their understanding of the rationale and reasons for treatment, such as prosthetic fitting will help ease the child's progress. Members of the family we sometimes call the SOG or "silly old grandparents" are very influential and it is extremely important to include them in the orientation and subsequent treatment planning.

The treatment programme for the limb deficient child must be "home centred". Except for surgery, treatment should be as far as possible on an outpatient basis. Prosthetic fitting and subsequent training in the use of the device need not require hospitalisation. In fact, there are three good reasons why hospitalisation should be avoided. The first is that hospitalisation implies the patient has an illness which requires medical attention in a hospital setting with the attendant hospital gowns, frequent measurement of vital signs, blood tests, nurses, etc. Our patients are not "sick". Secondly, they are being provided with an artificial limb, followed by therapy. This can be done as an "out-patient". Patients/family coming from a distance can be housed in guest rooms, friend's/relative's homes or motels. The use of outside housing usually means the costs are much less, except that often insurance and governmental agencies will not cover non-hospital expenses. The third and most important reason for out-patient treatment is that carry-over of the treatment programme can be accomplished better. Out-patient prosthetic fitting and treatment requires one or both parents or carers to bring the child to the clinic. They can be included in the fabrication and therapy process and have a better appreciation of the abilities of their child relative to the prosthetic programme. They are then better prepared to encourage the child to use the prosthesis in the home situation. They can explain the functions of the prosthesis and its benefits to other family members, neighbours and school personnel.

In order that the treatment programme be a comprehensive one for the limb deficient child, a co-ordinated effort is required to involve the local physician, local treatment (O.T., P.T., prosthetics) agencies, school, friends, and community programmes such as clubs, churches and employment.

One of the most critical issues in the treatment programme especially in the USA where we do not have some type of socialised medicine or catastrophic health insurance, is the family's abilities to afford the costs of medical care for the limb deficient child.

The cost of prostheses in the past 10 years has increased a great deal because of the number of "high technology" prosthetic devices now available.

In the USA the cost of medical care (including physician, therapists and social worker's fees, x-rays, surgeries and prosthetic costs) are covered in one of four ways. If the family has sufficient resources, they may manage all costs themselves. However, this is rare. Approximately 20–40% of our patients have private insurance (either standard health insurances with deductible payment by the insuree or Health Maintenance Organisations through contracted groups). Government assistance programmes such as Medi-Care

(Medi-Cal in California) and Crippled Children's Services (a programme established by the Federal Government in the 1930's to assist families with orthopaedically and/or physically handicapped children with contributions towards the cost of medical care from the county, state and federal agencies), will provide funds for families who qualify financially (earnings under $40,000/year) and whose child has a medically eligible condition. The final method of payment for treatment comes from private philanthropic groups such as City of Hope, Shriners Hospital for Crippled Children and Variety Clubs.

The problems with all of these funding programmes is that there is little consistency in the assistance to the families. Factors such as: pre-existing condition, i.e. the condition existed before insurance coverage started; extent and type of coverage, frequency of payment for durable goods are so different that families are never sure that financial coverage of care is available. Even with the same insurance companies, the actual coverage for prosthetic care varies. In some cases externally powered prostheses, and the more expensive lower limb "hi-tech" devices such as Endolite and S–N–S systems are covered for some, but for other families are refused. In cases of infants, some companies only pay for the first prosthesis and not for subsequent replacement even though replacement is necessary due to normal physical growth. Finally, the cost of prostheses, especially the more expensive ones, varies as much as $2,000, depending on the location in US and even among shops in the same area.

Therefore in prescribing prostheses the clinic must not only concern itself with the medical condition and the needs of the patient, but also whether the cost of the system or components can be paid for by the family or its third party payer. Often this decision is not up to the clinic but to the payer of the prosthetic or rehabilitation programme.

A major concern in most clinics in the US is who should make the final decision regarding prosthetic prescription. In countries such as the UK, Germany and Japan, the clinic team decides, based on criteria developed by the team. In the US, the family through its insurance or payer can often dictate prosthetic treatment even if the team is *not* convinced that the treatment programme is the best. Hopefully we can come to some objective criteria that will apply to all patients regardless of their financial status.

Summary

The team approach is considered the optimal to the care of the limb deficient child. A co-ordinated programme utilising the expertise of experienced personnel and taking into consideration the developmental needs of the child will allow the child to grow and function as normally as possible. The goal of the team is to assist the patient to become a productive, self-sufficient member of society.

Prosthetics and Orthotics International, 1991, 15, 82–87

The prosthetic treatment of upper limb deficiency

B. CURRAN and R. HAMBREY

Disablement Services Centre, Withington Hospital, Manchester, UK

Abstract

The provision of prostheses for children with upper limb deficiency starts at an early age. The roles of the occupational therapist and prosthetist in this programme are described. Casting and fitting techniques applicable to transverse forearm loss and aspects of myoelectric hand prostheses are included. An outline of training methods as related to both parent and child is explained. Higher levels of limb deficiency are covered briefly and the use of alternative methods of electric hand control is included. Finally, a brief summary of distal deficiency is discussed.

Introduction

The treatment of children with congenital limb deficiencies is carried out at this centre by a team comprising doctors, prosthetists and an occupational therapist. A social worker is also available for counselling when necessary. Early referral is encouraged so that the parents can be helped as soon as possible.

Transverse forearm deficiency

A first prosthesis can be fitted as early as two months of age. It consists of a plastic socket to accommodate the stump, bonded into a foam-filled cosmetic glove (Fig. 1a). The provision of such a prosthesis:

1. meets the need of parents for cosmetic replacement;
2. provides the child with an early experience of wearing a prosthesis;
3. lengthens the deficient limb and provides passive assistance to the sound hand;
4. assists the child's balance.

The socket of the first prosthesis can do little more than contain the volume of the stump. It should be thin-walled so that it may be stretched to accommodate growth whilst a new prosthesis is being manufactured. Progress is reviewed at two to three month intervals, or more frequently if the parents' wish. The child will continue to wear a passive prosthesis until aged fourteen to eighteen months, when consideration is given to providing a more functional type consisting of a self-suspending socket, with activation of a terminal device by a cord and single shoulder loop. Two alternative devices are available — a small lightweight split hook and the CAPP terminal device (Figs. 1b and 1c). For the past nine years the CAPP device has been preferred because:

1. it is inherently safer than the split hook, and therefore can be fitted sooner, enabling the child to develop early awareness of its functional benefits;
2. it has a more efficient gripping surface than the split hook;
3. the opening span is wide, enabling a small child to grasp things easily even though co-ordination is not well developed;

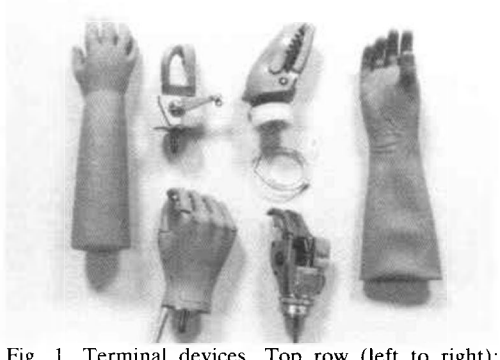


Fig. 1. Terminal devices. Top row (left to right): (a) Cosmetic foam filled glove. (b) Lightweight split hook with plastic covered jaws. (c) CAPP terminal device. (d) Cosmetic cover for hands. Bottom row (left to right): (e) Child's mechanical hand (cable operated). (f) 2 inch myoelectric hand.

All correspondence to be addressed to B. Curran, DSC, Withington Hospital, Cavendish Road, Manchester M20 8LB, UK.

4. the appearance has been found to be more acceptable to parents;
5. it has been found that less formal training is required (as skills seem to develop more naturally and automatically).

If the CAPP device is rejected the child and parents are encouraged to continue with the cosmetic prosthesis, which may be provided in addition to the functional arm for social reasons.

Transverse upper arm deficiency

The first prosthesis is provided at six to nine months of age for this level of deficiency and is similar to that provided for forearm loss, i.e. a laminated socket bonded in a flexed position to a foam-filled forearm and hand. Suspension is via a web harness which later provides attachment for control cords. As the child develops, an elbow joint is required and flexion provided via bi-scapular excursion. Once this is achieved with conviction, an elbow lock can be added, thereby allowing the flexion cord to be utilised for operation of the terminal device. As with the forearm levels, the CAPP device is preferred and is fitted once the child has gained control of elbow flexion and locking.

The occupational therapist's (OT) role initially is to advise on wearing the prosthesis, to reassure the parents about the child's development and to give information about help available from other professionals and voluntary bodies.

When a functional prosthesis is provided the OT spends more time with parent and child to familiarise them with the arm, albeit in an unstructured manner. It is imperative that parents are aware of their child's development, both physically and mentally, so that their expectations of achievements with the prosthesis are realistic. If the child does not appear to be making progress with the CAPP device 2–3 months after delivery, structured training sessions are arranged. In these, activities are chosen to encourage awareness of the use of the prosthesis, with passive assistance by the OT initially when necessary.

The spring which controls the grip force of the CAPP device is available in two strengths, the weaker one being used until the child is competent. Further sessions may take place if prosthetic use does not increase in line with normal development. At all times the parents are advised how to help the child develop skills with the prosthesis.

The provision of an electrically powered hand is usually considered between the ages of three and four years before starting primary school (Fig. 1f).

This type of hand provides good prehension with the minimum of physical effort, combined with good cosmetic appearance. It is necessary to give parents accurate information because they may have unrealistic expectations acquired from the publicity given to these hands, and it is important to determine the optimum age for fitting. Consideration is given to the child's ability to accept a new type of prosthesis, his attitude to his existing prosthesis, his level of co-operation and also the attitudes of his parents. Physical factors, such as the length and shape of the stump, the ability to produce good myoelectric signals and the stature of the child, play a part in the decision. Myoelectric control is the preferred option for transverse forearm cases.

An alternative method of hand control is a "servo" system, in which an electronic circuit causes the hand to move in synchrony with an external transducer thus providing positional feedback. This has several advantages over myoelectric systems for those who are prepared to accept minimal harnessing and an external lead, and is especially appropriate when it is difficult to locate controllable myoelectric signals, or when there is a problem in providing the required accuracy of socket fitting. Another advantage is that the suspension and control

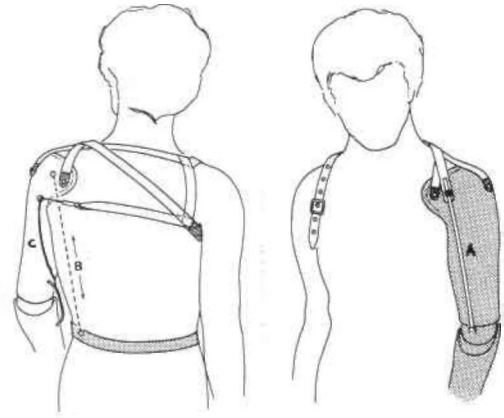


Fig. 2. Typical arrangement of control cords for above-elbow prosthesis. (a) Elbow locking. (b) Hand operation (servo). (c) Elbow flexion.

techniques used in body-powered prostheses are quite adequate to control the servo hand. The only major differences are that the forces and excursions required for control are small and that operation of the hand is possible with less discomfort, exertion or gross body movement.

In some cases the child may change from servo to myoelectric control if, through growth and maturity, better signals are eventually achieved. For transverse forearm deficiencies a minimum amount of harness is required to control the servo hand, i.e. a shoulder loop, but for upper arm cases a full harness and suspension are needed for elbow flexion and locking, as well as hand control (Fig. 2).

Prosthetic techniques

Below-elbow myoelectric prostheses may be retained by any suspension technique as long as the electrodes remain in continuous contact without slippage under normal loading conditions and at all angles of elbow flexion. One of the major benefits of below-elbow myoelectric systems is that the prosthesis can be made to be self-suspending because body activation of terminal devices is no longer necessary. Although self-suspending sockets have been available for some time, earlier prosthetic methods limited flexion quite severely and were therefore not as well accepted as they might have been. These sockets relied on a reduced antero-posterior measurement to provide suspension over the olecranon. The newer techniques such as those developed in the UK and elsewhere rely on a decreased mediolateral measurement above the condyles of the humerus, thus freeing the anteroposterior dimension to give the maximum range of flexion and extension.

Two casting techniques are used and are typical of modern prosthetic practice. The first relies on moulding the plaster bandage over the condylar areas whilst the cast is still wet, thus forming suspension areas. The second involves no primary moulding. Instead, a suspension "brim" is formed by rectification of the positive plaster model. The only criterion as to the choice of technique is that the prosthetist should feel confident enough with it to provide a comfortable and reliable socket. In order to ensure good contact it is desirable that the electrodes are held firmly against the skin during all normal loading conditions. They should produce distinct marks on the stump which show all the features of the electrode face. However, they should not indent the skin so deeply that discomfort is caused. Care should be taken to avoid sites over bony areas or scar tissue. If distinct marks are not found, the depth of the electrode position should be increased or the electrode resited to improve contact. Electrode positions which give high signal response on the myo-tester may be identified on the posterior aspects of the medial and lateral stump surfaces. It is not advisable to site the electrodes towards the posterior aspect of the stump as it is likely that contact will be lost during part of the flexion range. A trial fitting prior to completion of the prosthesis is an essential feature, and enables the prosthetist to assess the accuracy with which the definitive prosthesis will match the original intentions at the plaster cast stage. An inner socket is manufactured from the cast and contains all the features necessary for suspension and electrode placement. At this stage a set of equipment — the hand, cables, battery, electrodes etc, as well as a temporary outer socket must be available. The latter can be made from a laminated cylinder with an internal diameter which accepts a friction wrist housing at one end (Fig. 3). The use of a transparent thermoform socket has been found to be extremely useful when refining technique. Not only can the prosthetist identify areas of the socket which are inappropriately loaded, but he can also locally modify these areas with the careful use of a heat gun. The initial stage of fitting is concerned with socket comfort and retention. After removal from the cast the socket is trimmed to remove surplus material and sharp edges are smoothed with sandpaper. The temporary outer socket is aligned on the check socket and

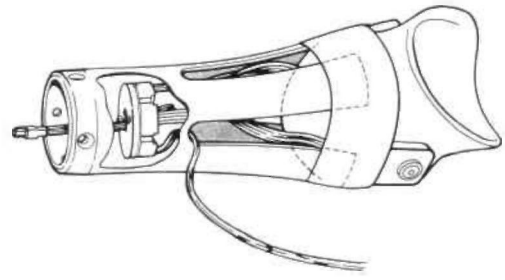


Fig. 3. Temporary prosthesis for myoelectric fitting.

held in position with adhesive tape. Length is not a major consideration at this point, although clearly some approximation to the required length is desirable. The temporary prosthesis, complete with hand is then fitted to the child. Comfort, alignment and suspension are now rechecked and adjustments made where necessary.

The fitting procedure now moves on to the next stage which tests the electrode placement and the operation of the hand. It is essential that the child acquires a basic understanding of the control of the hand before the prosthesis is completed.

It is believed that the fitting stage is the time to carry out any rectification of the socket and alignment, thus ensuring as far as possible a trouble-free acceptance by the child when the prosthesis is delivered and training commences.

Occupational therapy

It has been found that immediate training in the control of a myoelectric hand is an essential part of provision, especially with young children. If this is not available a few *may* learn to control it by trial and error, but many will be unable to establish adequate control quickly and consistently enough to prevent frustration and disillusionment. During this early training it is vital to spend time establishing a balance between the settings of the opening and closing electrodes, in order to ensure that success is quickly achieved. The child is seen by the OT at the casting and the fitting stages. This enables her to make observations of the child's ability prior to the prosthesis being completed.

Following delivery the child attends the OT department for two days basic training, during which emphasis is placed on using the prosthesis in bilateral functional activities. A prosthetist is available for any adjustments to be made during this time. Further training may be given 1–2 weeks later if required. Liaison with schoolteachers and others regularly involved in the child's care ensures that the prescribed regime is followed. This is particularly important in order to prevent undue pressure being put on the child which may contribute to rejection. The amount of training given often depends on the level of use of the previous prosthesis. A review takes place after 2–3 months but once well established, intervals between appointments may lengthen. Responsibility is then with the parents to notify the centre when problems occur. With the servo hand very little training is needed from the point of view of hand control, but the parents should be instructed in the care of the equipment and may still need to adjust their expectations of the prosthesis. In higher levels of loss such as upper arm and total shoulder deficiency, time will be spent on achieving control of a functional elbow unit when this is fitted.

Bilateral limb deficiency

The treatment of children with bilateral deficiencies requires even closer co-operation between the professionals involved. As these deficiencies are rare it is considered that the children should attend specialist centres where the staff have built up considerable experience over a number of years. Although no two children are exactly alike the problems presented have a common core and experience gained with one child can be of considerable assistance in dealing with another. Psychological and practical support is essential for parents from the earliest possible time to help them to adjust and create the right environment in which to rear the child. The aim is to enable the child to see himself as having the least handicap. This can be achieved by a balanced combination of prosthetic provision and continuous assessment of ability by an occupational therapist.

It has been found, especially in the more proximal deficiencies, that children often achieve greater independence without prostheses. This does not mean that they should not be fitted because it is important that the child and parents see the provision of prostheses as a part of a total programme of treatment. Early experience of prostheses will enable the child to make a more effective choice at some future date.

Although a programme of prosthetic treatment may be mapped out for the first few years, invariably this is changed because it is impossible to forecast progress accurately, both in the development of the child and that of available components. For total shoulder and upper arm deficiencies, both sides are fitted with prostheses from about the age of 6–12 months. These prostheses consist of sockets with friction elbow units and foam-filled

forearms and hands made in one piece. The children are not expected to wear the prostheses for long periods but they fulfil certain requirements for the developing child and parents. These are as follows:

1. preparing the child for probable future prostheses;
2. creating early involvement of parents in the programme;
3. providing simple functional ability.

The main purpose of occupational therapy is to enable the child to reach the highest level of independence both with and without prostheses. Maximum use of residual limbs, however small, should be encouraged and much time will be spent developing use of the feet.

Feeding will be the first functional daily activity to be developed, followed by dressing, writing skills, toiletting etc, as appropriate to normal child development. Aids will need to be provided but these should be kept to a minimum and as simple as possible. They should also be easily portable to ensure independence in any location.

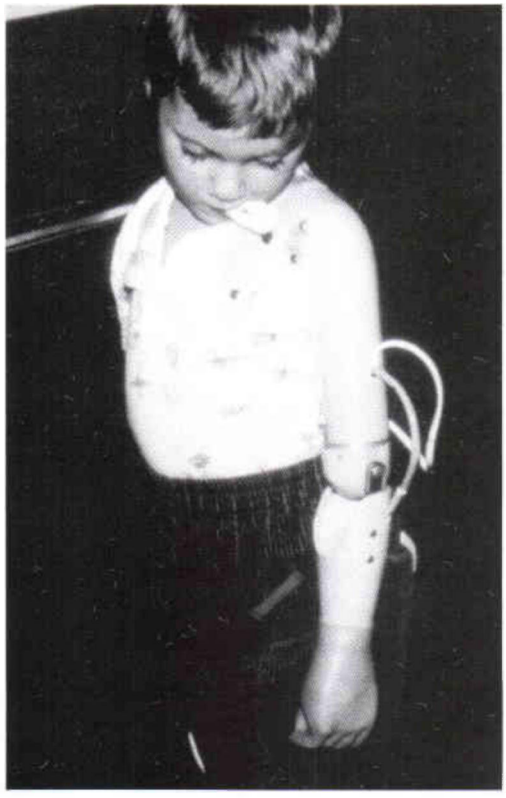


Fig. 4. Prosthesis for bilateral total upper arm deficiency. Note — voluntary elbow flexion, bite operated elbow lock and electric hand.

Prosthetic training is given in short sessions as and when necessary and with close co-operation with community-based paediatric OTs. Activities to develop skills with the various different mechanisms should be purposeful and appropriate to the age of the child and advice is given to the parents about encouraging the child to develop spontaneous use. Meeting other children can be helpful in learning by example.

From the first simple prostheses the child will progress on to more complex elbow mechanisms with the ability to lock the elbow in varying degrees of active flexion, and a terminal device fitted from the range already described. "Bite tabs" are used for elbow locking when the child is unable to operate a standard mechanism. Consideration is given to electrically powered prostheses along the lines already described for unilateral deficiencies (Fig. 4).

Distal deficiency

Distal longitudinal and transverse deficiencies pose a different problem in the sense that replacement of function takes priority over that of cosmesis in the early years, although the reverse may happen later. If a carpo-metacarpal joint is present, the rudimentary palm can oppose to a shaped plate mounted on a forearm gauntlet proximal to the wrist. Such a device may be made of plastic, leather or wire. The development of a simple plastic-coated wire device for holding a knife, fork or spoon has proved extremely successful for children (Fig. 5). By the very nature of

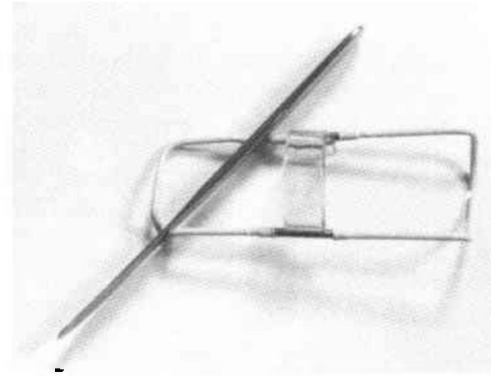
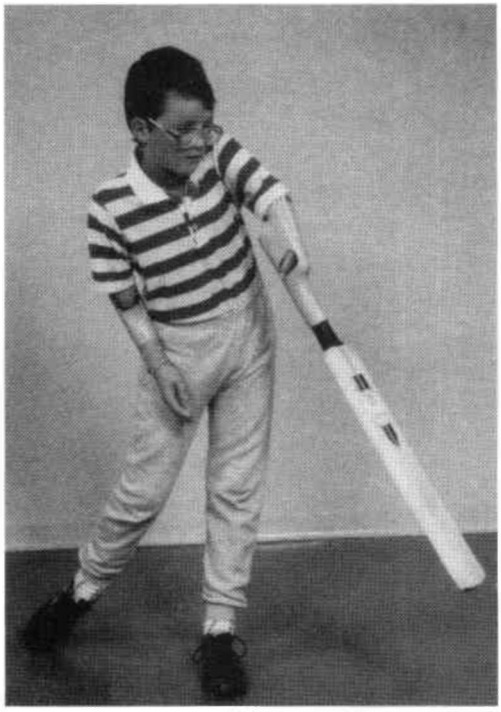
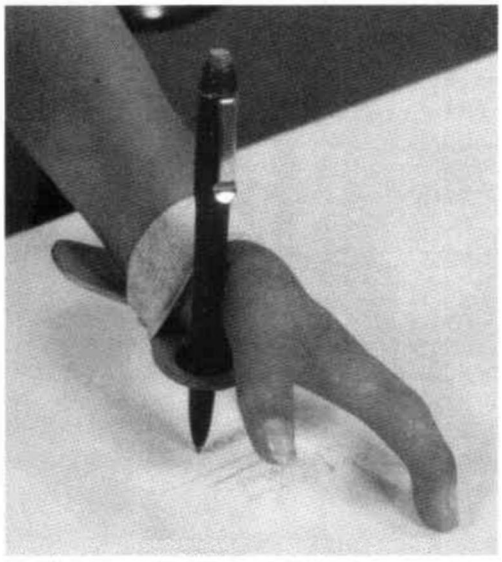


Fig. 5. Wire opposition device to hold knife or fork for hand deficiency.



(a)



(b)

Fig. 6. Opposition devices. (a) To hold cricket bat. (b) To hold pencil.

distal deficiencies opposition plates have to be designed individually according to the function required and that already available. Much ingenuity is required to design simple yet effective devices which can be provided for a variety of everyday activities — educational, social and recreational (Fig. 6). Cycling, holding a skipping rope, writing and holding musical instruments are examples. In some cases the OT may solve the problems by the provision of external aids and a wide range of these is available, such as adapted scissors, non-slip mats, special cutlery, and writing aids. Every encouragement should be given to enable the child to use the deficient hand in a normal manner.

Children's needs are changing constantly throughout childhood, and correspondingly their prosthetic requirements also change. Sometimes children choose not to wear a prosthesis for a period of time, but this can change very suddenly, so they should be reviewed in the centre periodically even when in "a non-wearing phase". Parents should always have access to the professionals when required.

Because the replacement of a hand is so complex, there is frequently a need to have two different types of prostheses at the same time to suit the varying activities in which children are engaged. No direct charge is made to the client for any limb prostheses provided by the National Health Service in the UK even though there is a higher cost in providing electrically powered prostheses. Thus no pressure is brought to bear on any member of the clinic team when selecting the most appropriate prosthesis for the child. It is essential for doctors, prosthetists and therapists to keep abreast of new developments both in hardware and techniques, in order to provide the best possible service to these children.

REFERENCES

ATKINS D. J., MEIER R. H. (1988). Comprehensive Management of the Upper Limb Amputee, Springer-Verlag.

HAMBREY R., WITHINSHAW G. (1990). Electrically powered upper limb prostheses: their development and application. *Br. J. Occup. Ther.* **53**, 7–11.

Steeper Electric Hand — myoelectric and servo systems for the Steeper electric hand: *technical manual*. London: Hugh Steeper Ltd.

DAY H. J. B. (1988). The prosthetic treatment of congenital upper limb deficiency. In: *Amputation Surgery and Lower Limb Prosthetics*. Edited by G. Murdoch. Oxford: *Blackwell Scientific*, p306–311.

Prosthetics and Orthotics International, 1991, 15, 88–92

Structured training of children fitted with myoelectric prostheses

L. M. HERMANSSON

The Arm Prosthetic Clinic, Örebro Medical Center, Örebro, Sweden.

Abstract

This paper presents an occupational therapy method for training children fitted with myoelectric prostheses. It is based upon a structured way of describing the accomplishments of a child fitted with a myoelectric prosthesis, called the Skill Index Ranking Scale (SIRS).

By using the SIRS when designing the training session, the therapist can progressively increase the demands presented to the child. Furthermore, the SIRS facilitates for the therapist the documentation and communication of the childs ability with the myoelectric prosthesis.

Introduction

Occupational therapy for children with myoelectric prostheses started developing after the successful fitting of a preschool child in 1971, (Sörbye, 1977). According to Sörbye *et al.*, (1978) myoelectric prostheses for children above three years of age have been available since 1976.

The facility founded by Dr. Rolf Sörbye in the Örebro Medical Center (ÖMC) is Sweden's major centre for childrens upper-limb prosthetics. Some 75% of 110 children supplied with myoelectric prostheses according to the Sörbye concept (Sörbye, 1977) are using their prostheses all day (6 hours or more, 4–7 days/ week). Out of the 110 children, 11% are using the prostheses half the day (2–5 hours 4–7 days/ week), and 6% are using it for a certain task at least once a week. The rest are using it sporadically (6%) or never (2%). About 10–15 new children are referred to the clinic each year.

All correspondence to be addressed to Liselotte Hermansson OT, Dept of Clinical Neurophysiology, Örebro Medical Center, S–701 85 Örebro, Sweden.

As patients are referred to ÖMC from all of Sweden, the more frequent prosthetic training is transferred to local teams. A local occupational therapist (OT), selected for the once-a-week training with the child is initially contacted by the OT at the clinic at ÖMC. The local therapist is invited to a 2-day visit in Örebro to learn about the prosthesis and the training programme. Since 1989, ÖMC has run courses for further education of OTs involved in the training of children with upper-limb prostheses.

Occupational therapy aims that the child should achieve age-appropriate independence and be able to choose to perform whatever activities he wants. The child should be as bi-manual as possible in performing the activities of daily living (ADL), the prosthesis being used as a normal, non-dominant hand. The training attempts to help the child to: 1) wear the prostheses, and 2) use the myoelectric hand.

The procedure of prosthetic fitting and training has been described earlier by Richardson and Lund (1959), Shaperman (1960[1,2]), Clarke and Patton (1980), Agnew and Shannon (1981), Marquardt (1981), Challenor *et al.*, (1982), and Garza (1986). In this paper a structured method to describe the accomplishments of a child fitted with a myoelectric prosthesis is presented. The method may be a tool for therapists in organizing training and to document and communicate a child's ability.

The Skill Index Ranking Scale (SIRS)

To provide a description of children's ability with myoelectric prostheses, a SIRS has been developed at the Arm Prosthetic Clinic at ÖMC (Table 1). According to this scale, the child's accomplishments with the prosthesis may be described on a 14 step scale. Every step on this

scale puts a higher demand on the child, compared to the previous step. By attending to the 14 step scale during training sessions the child is always presented with a new challenge. The steps represent different levels of function when using an electrical hand-prosthesis.

The SIRS is based upon:

- Sollerman's (1980) classification of grip types;
- the clinical occupational analysis as described by Hobbs-Cubie (1985);
- 20 years of clinical experience of developing skills among children with myoelectrical hand prostheses.

Principles for training

To be successful in training, the parent's support is vital. They are the ones who have to meet the day to day demands and support the child through trials and errors. The OT co-operates closely with the parents, and shares methods with them. The Swedish Dysmaelic Society, founded in 1979 on the initiative of Dr. Sörbye, supports the parents on social, financial and medical issues. Children with both/either upper and lower limbs affected, either through congenital malformation or other causes are, together with the entire family, members of the society. There are two meetings held each year, with a large number of attendees. Representatives from ÖMC regularly attend those meetings for consultation and to see the patients fitted in Örebro.

Table 1. The Skill Index Ranking Scale

1. Wear the prosthesis.
2. Use the prosthesis as support.
3. Spontaneously move and preposition the amputated prosthetic side.
4. Spontaneously place the terminal device (TD) in position and use it for support.
5. Control the grasp – release function of the TD.
6. Use a transverse volar grip, with the weight of the prosthetic limb unloaded from the child.
7. Use the tripod pinch, still without the weight of the prosthetic limb.
8. Use the transverse volar grip, without support for the prosthetic limb.
9. Use the tripod pinch with no support for the weight of the prosthesis.
10. Control the grip in various positions around the body.
11. Manipulate objects by changing their position in the TD.
12. Adjust the grip force in the TD, i.e. to hold without damage.
13. Control the grip with the arm moving, i.e. throw things with the arm hanging down.
14. Control the grip while moving the arm, throw things from above the shoulder.

Before the training commences, the OT observes the child to determine the current SIRS level. Thereafter the OT may choose an activity (according to the occupational analysis as described by Hobbs-Cubie (1985)) that increases the demands and permits the child to improve its ability. The activities chosen shall be age-appropriate. Training may be carried out on more than one SIRS level at a time. For example, a child at level two may perform grip-training while, at the same time, spontaneity is aimed at. The training must always be fun and encourage prosthetic use. The training in Örebro is mostly performed in individual sessions, when the OT is alone with the child. The relationship between the OT and the child is very important. Parents may be present until the child feels safe and secure with the therapist. Group sessions may be arranged depending on the number of children in a particular area. In a group of children with similar prostheses, problems and abilities, the children get support and help to further develop their abilities. After the training sessions the parents are told the current status and achievement. Videotape recordings are used to show the child's ability, and they sometimes get "homework" to practise until the next training session.

A very important factor for successful prosthetic fitting, is the follow-up in Örebro. Every six months if the family has not been in contact with the clinic, the OT calls them to check how the child is doing, and if everything is working as planned. An adequate prosthetic maintenance service is provided as well as spare prostheses for the child.

To wear the prostheses (level 1–4)

In order to stimulate pre-school children in prosthetic use, the therapist first may have to divert the interest of the child from the prosthesis, and its weight, using interesting objects and/or games. Thereafter the child may be made aware of the usefulness of the artificial limb, through some activity, and finally get credit for using it. For the older child, wearing should first be connected to certain activities and/or situations. When the habit of wearing the prosthesis is established, a wearing-habit that, finally covers most of the day is aimed at.

If the prosthesis seems to obstruct the movements of the child, the OT must guide the

child into a better movement pattern. As the prosthesis must be regarded as an extension of the residual limb, it should not impede but improve the limb's function. It should be involved in the childs body-image and, for example, swing naturally when walking, and be placed spontaneously on the table when sitting.

Training through bilateral gross motor activities for the arm, combined with wearing-training may, in time, make it easier for the child to obtain spontaneity and supportive use of the prosthesis.

The first four steps in the SIRS may be used for documentation of skills with a passive Terminal Device (TD), and as criteria for when to change to an active TD as well as for training with a myoelectric prosthesis.

To use the myoelectric hand (level 5–14)

For children, with previous experience of a passive TD, movements in the myoelectric TD appear spontaneously and involuntarily. Through visual feedback the child learns to control opening/closing in a reasonably short time (between one day and a fortnight). Difficulties in controlling the myoelectric hand sometimes, in the experience at Örebro, occur among children fitted when older than five years of age. It seems to be the same problem as adults have when first fitted with a myoelectric hand. To find the right muscles for contraction, biofeedback visualized on a screen where muscle activity may be separated, is sometimes used. Biofeedback is used in the same way when there are problems with co-contraction (i.e. when antagonists are activated simultaneously with the agonist).

The child may find the myoelectric prosthesis, heavy when first fitted and this may interfere with the ability to control the grip. The SIRS suggests, therefore, that the training commences with activities where the weight of the prosthetic limb is unloaded from the child. For example sitting with the prosthetic arm on the table, holding an object in the TD and working upon it with the other hand. As the child's ability increases, the difficulty of the task increases. Throwing things from above the shoulder is the most difficult thing to do.

As its prehensile skill increases, the child will be able to use the artificial grip more effectively. When manipulatory skill is achieved, the child will learn to adjust the grip force to the object, i.e. to hold hard enough without damaging the object. Sollerman (1980) describes eight different grip types. The most commonly used electrical prosthesis offers two grip types, the transverse volar grip and the tripod pinch. It may, however, be possible to use the myo-hand for the other grip types as well, although not quite so well.

Clinical experience of children with myoelectric prostheses has shown that some of them have been able to learn to control the grip, before actually having achieved spontaneity and supportive use. They do not, however, use their prostheses in daily life and the usefulness of the prostheses is therefore small. Ability at all levels of the SIRS is needed if optimal use of this sophisticated tool is to be obtained.

Intensive training camps

Since 1978, intensive training has been used to treat children fitted with hand prostheses. Each year during the summer holiday, about one week before school begins, children, 6–9 years of age, accompanied by one of the their parents, gather to attend the training camp.

Twice during the week the physician and prosthetist visit the camp for examinations and prosthetic maintenance.

The aims of the training camp are that the child will be able to use the prosthesis spontaneously in daily life, and that the parent will be able to support the child in its development.

The intensive training camp attempts to:
- support the child in developing a positive self-image;
- motivate the child to prosthetic use;
- support the child in developing bimanual skills;
- support, teach and guide parents.

The intensive training camp features:
- a structured programme based on play-activities;
- demanding, fine motor activities in the morning;
- social and gross motor activities in the afternoon;
- age and ability related groups;
- parental participation in training;
- parental discussions;
- closing festivities.

To know the child's prosthetic use and ability, the OT initially interviews and assesses the child (according to the SIRS), and forms an individual programme. The assessment is repeated at the end of the week to evaluate the result.

The results from a training camp show that the attendees get increased prosthetic use as well as use of the artificial grip. Furthermore, the children get better self-confidence.

Training with multifunctional and bilateral prostheses

Multifunctional electrical prostheses have been supplied to a few cases at ÖMC. These prostheses normally feature a powered TD and a powered elbow or wrist unit. In order to evaluate the possibilities for a patient to operate a multifunctional prosthesis, a computer based control system has been developed (Philipson, 1985; Philipson and Sörbye, 1987).

By means of this device it is possible to design a circuit that controls one to eight different output states from two myoelectric electrodes. The benefits of this system are that all parameters of the control (i.e. sensitivity, switch levels, degree of acceptable co-activation etc.) may be easily adjusted in the software. With the device, it is also possible to find out whether or not the patient is a possible candidate for multiple-state control.

The computer based myoelectric control system has proven to be a useful tool for training and evaluation of patients. It also saves time since a complicated prosthesis does not have to be built until the patient has shown the capacity to operate it.

In cases of bilateral deficiencies the children at ÖMC usually get a powered prosthesis on one side and, in the case of a high level deficiency, a passive or body powered prosthesis on the other side. The training follows the SIRS, with sufficient adaptations with respect to the limitations imposed by the deficiency.

Financial implications for myoelectric prosthetic fitting

In Sweden, medical care is provided by the tax financed social security system. This gives the parents different kinds of economic support (Fig. 1).

A patient pays a small fee to see a doctor in Sweden. If the orthopaedic consultant considers that the child would benefit from a prosthesis, all the equipment is free of charge. That includes one, or more, prostheses, at least three batteries and a battery-charger. Furthermore, new gloves, change of socket and repair is free. Training and follow-up are, financially, regarded in the same way as the medical consultation, i.e. associated with a small cost.

Child care allowance

If you have a handicapped or a sick child who needs special supervision and care for at least six months, you can obtain a child care allowance. Additional costs that you incur are also relevant when the insurance office decides whether you are entitled to a child care allowance. The child must be under the age of 16 years. It the child is cared for in an institution you can obtain a child care allowance for the days when the child is at home. If the child needs a great deal of supervision and care, you can obtain full child care allowance. There is also a half child care allowance (and as from 1 July 1988 also a quarter allowance). If there are several handicapped children in the same family the insurance office will consider the total need for supervision and care and the additional costs. This means that a child care allowance may be obtainable even though none of the children would qualify individually for such an allowance.

Part of the child care allowance may be exempt from tax.

For further information, read the brochure **Child Care Allowance and Disability Allowance.** Obtainable from the social insurance office!

From "Useful information on Social Security"
Published by the Federation of Social Insurance Offices FKF 79–1 88.04 BG.

Fig. 1. Part of the financial implications for children with limb deficiencies in Sweden.

Conclusion

The training of children with myoelectric prostheses according to the SIRS has shown to be an efficient way to achieve a high level of independence. In Örebro, the SIRS is an integrated part of the entire fitting and training procedure. The scale has proved to be a useful way of describing the current skill level of an amputee. It is used to organize the training sessions effectively and also for assessment of the child prior to a change from a passive to a myoelectric terminal device. Furthermore, the SIRS can be used as an evaluation tool when studying which training technique is the most efficient.

Acknowledgements

This project has been made possible by the financial support of the Research Committee at Örebro County Council.

The author would like to express her gratitude to Lennart Philipson PhD, Kristina Waldenlöv OT and Pehr E. Leissner MD for guidance and support throughout this study.

REFERENCES

AGNEW, P. J., SHANNON, G. F. (1981). Training program for a myo-electrically controlled prosthesis with sensory feedback system. *Am. J. Occup. Ther.*, **35**, 722–727.

CHALLENOR, Y. B., RANGASWAMY, L., KATZ, J. F. (1982). Limb deficiency in infancy and childhood. In: The child with disabling illness. Downey, J. A., Low, N. L. (eds). — New York, Raven Press. p. 409–447.

CLARKE, S. D., PATTON, J. G. (1980). Occupational therapy for the limb-deficient child. *Clin. Orthop.*, **148**, 47–54.

GARZA, P. (1986). Case report: occupational therapy with a traumatic bilateral shoulder disarticulation amputee. *Am. J. Occup. Ther.*, **40**, 194–198.

HOBBS-CUBIE, S. (1985). Occupational analysis. In: A Model of Human Occupation. Kielhofner, G. (ed) — Baltimore, Williams & Wilkins. 147–155.

MARQUARDT, E. (1981). Prothesen für armlose Kinder und Jugendliche — Erfahrungen und Folgerungen (Prostheses for armless children and young people — experiences and conclusions). *Rehabilitation*, **20**, 33–41.

PHILIPSON, L. (1985). Adaptable myoelectric prosthetic control with functional visual feedback using microprocessor techniques. *Med. Biol. Eng. Comput.* **23**, 8–14.

PHILIPSON, L., SÖRBYE, R. (1987). Control accuracy and response time in multiple-state myoelectric control of upper-limb prostheses. Initial results in nondisabled volunteers. *Med. Biol. Eng. Comput.*, **25**, 289–293.

RICHARDSON, G., LUND, A. (1959). Upper extremity prosthetic training for the young amputee. *Am. J. Occup. Ther.*, **13**, 57–63.

SHAPERMAN, J. W. (1960[1]). Orientation to prosthesis use for the child amputee. *Am. J. Occup. Ther.*, **14**, 17–26.

SHAPERMAN, J. W. (1960[2]). Learning techniques applied to prehension. *Am. J. Occup. Ther.*, **14**, 70–74.

SOLLERMAN, C. (1980). The use of eight main handgrips in activities of daily living. In: Grip function of the hand. Analysis, evaluation and a new test method, (PhD thesis) Sollerman, C.-Göteborg, Gotab. p1–14.

SÖRBYE, R. (1977). Myoelectric controlled hand prostheses in children. *Int. J. Rehabil. Res.*, **1**, 15–25.

SÖRBYE, R., HEDSTROM, L., HOLMQUIST, T., RANDSTORM, S. (1978). Development and improvements of a myoelectric controlled hand prosthesis for children, based on clinical experiencies. In: Advances in External Control of Human Extremities; Proceedings of the Sixth International Symposium on External Control of Human Extremities, August 28 — September 1, 1978, Dubrovnik. — Belgrade: Yugoslav Committee for Electronics and Automation. p475–480.

Prosthetics and Orthotics International, 1991, 15, 93–95

The use of electric elbows in the rehabilitation of children with upper limb deficiencies

W. F. SAUTER

The Hugh MacMillan Rehabilitation Centre, Toronto, Canada.

Abstract

For those with levels of loss above the elbow, the normal prescription choice is now considered to be the use of an electric elbow combined with a myoelectric hand using a single site three state control. The lightweight VASI unit controlled by a harness mounted switch is mounted in a prosthesis the socket of which is total contact, and allows improved power transfer.

Introduction

The use of electric elbows in upper limb prosthetics at the Hugh MacMillan Rehabilitation Centre (HMRC) dates back to when it was still known as The Ontario Crippled Children's Centre (OCCC).

In 1964 the staff of the Prosthetic Research and Training Unit (PRTU) at OCCC was charged with providing prosthetic and orthotic care for children referred to the institution's clinic.

Included in the 600 child amputees registered with the institution's clinic were 21 thalidomide children. These children had phocomelic limbs that lacked in reach, grasp and strength. It was the challenge of that group that triggered the team to experiment with externally powered components such as electric elbows, hooks and wrist rotators. A number of these children became adept at operating electric three-jaw-chuck terminal devices and elbows. Using the digits of their flippers to push against switches, some accomplished tasks such as feeding, picking up objects and even flicking on a light switch.

All correspondence to be addressed to W. F. Sauter, The Hugh MacMillan Rehabilitation Centre, 350 Rumsey Road, Toronto, Ontario M4G 1R8, Canada.

The experimental devices were primarily functional because it was believed that lack of function would lead to frustration and frustration to rejection. In retrospect, none of these patients became long-term users of their devices. Rejection can be directly attributed to the lack of cosmesis of the devices provided. Indeed, the acceptance and the success of externally powered prostheses began when aesthetically pleasing, functional hands became part of the prosthetist's armamentarium.

Patient selection criteria

In the past, the members of the team would sometimes discuss at length the pros and cons of conventional versus powered fittings. Today electrically powered components, wisely chosen, offer distinct benefits to the amputee. It is known that electric elbows offer more comfort through reduced harnessing and that they provide function to high level amputees when the alternative would be a cosmetic sleeve filler.

In HMRC the selection of patients for the provision of electrically powered prostheses has been reduced to agreeing not "if", but "when". The electrically powered prosthesis has become the conventional prosthesis.

Clinical experience with electric elbows at HMRC

From 1964–1968 several prototype elbows were field-tested on children and teenagers with upper limb deficiencies. The patient group included those with uni- and bilateral above-elbow loss as well as shoulder disarticulation and total arm deficiency.

Different modes of control were tried. These included mercury switches (attitude control),

arrangements of push and pull switches, and nudge control. Together with the elbows, various hooks, both cable and electrically powered, were evaluated for their usefulness as terminal devices.

In 1968, the final prototype elbow was passed on to Variety Abilities Systems, Inc. (VASI) for commercial production. To date, 600 of these units have been sold worldwide as the 62-type elbow. Compact and light, (370 gm) the 62-type elbow was found to be a rugged performer which has been used by HMRC in over 150 fittings, on adults as well as on children and teenagers. In some complex fittings involving bilateral, high-level adult patients, this elbow was not only used to provide forearm lift but found equally useful as a powered abductor or humeral rotator. In the case of elbow disarticulation patients, the VASI 62 elbow is the only elbow that can be used in a reversed position with the elbow becoming part of the forearm. Using this approach, the excess length of the humeral segment can be reduced to one inch (25 mm). While the elbow is normally supplied with a 6 volt motor VASI will supply the elbow with a 12 volt armature upon request. This model is about 25% faster but also needs a larger power pack.

From the many fittings carried out, a clear concept for an above-elbow prosthesis with electric elbow and hand developed. As a model it has remained unchanged to this day. In principle, the available myoelectric potential is reserved for control of the hand. Control of the elbow is kinetic, using extension of the humeral stump to trigger a pull switch. The patient is fitted with a "half-and-half" socket. The completed socket distal to the axilla shelf is made from rigid resin, whilst the shoulder portion is made from flexible silicone rubber laminate. The deltoid area is cut away. Anterior and posterior wings extend past the coracoid and scapular spine respectively. These sockets are almost self-suspending, show excellent rotational stability and most importantly, enable the patient to carry the weight of the prosthesis comfortably all day. These sockets are total contact but not suction sockets as it was found that neither children nor parents have the skill and determination to pull a stump correctly into a negative pressure socket. Suspension is aided by a single one inch (25 mm) wide transverse strap with velcro closure over the chest. The pull switch is mounted on the medial superior quadrant, of the socket's anterior surface so that it is floating and self-aligning and is fastened with a single screw. The cord or dacron strap is carried over the shoulder on the same side and fastened with a buckle to the transverse strap on the back. Thus, extension of the above-elbow stump will activate the switch and, consequently, the elbow. Involuntary activation is avoided because humeral extension is almost an unnatural movement. A single myoelectric control site for the hand can usually be found at the medial head of the biceps. However, especially in short above-elbow stumps, the medial head of biceps is not where it should be, but can be located more posteriorly and deeper into the axilla. In many years of prosthetic practice, only two patients at HMRC have been found to have signals from both biceps and triceps. The myoelectric control system for most above-elbow patients therefore, is a single-site 3-state system. HMRC have used the amplitude-sensitive VASI or UNB systems as well as the rate-sensitive Bock system. For small children, the so called St. Anthony's Circuit (cookie cruncher) has also been used. This is also a single-site system where the EMG signal causes the hand to open. When the patient relaxes, the hand closes automatically.

Unfortunately the batteries needed to drive the actuators and systems have not been designed to accommodate the prosthetist's needs. They are heavy and because of their shape and size, are difficult to place into the artificial limb. The batteries are placed as proximally as possible and if there is no space between elbow and socket, sometimes single button cells are spread on the outside of the socket and, after wiring is done, taped into place. The least satisfactory method is to have the battery pack attached to the harness which leads to endless cable and connector problems. It has also been seen that many patients and parents have problems with charging the batteries because the patient, not unlike a car driver whose gas gauge does not work, must judge the amount of charging needed.

About six years ago HMRC were asked to provide powered prostheses for a two-and-a-half year old girl with bilateral above-elbow amputations who was unable to obtain function

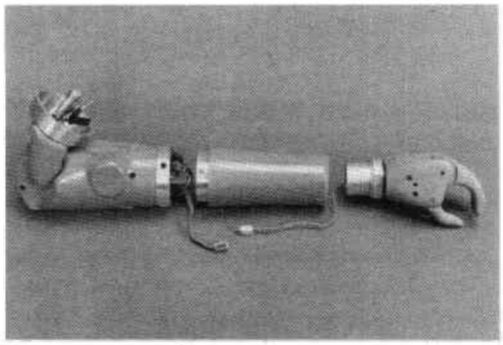


Fig. 1. The HMRC "mini" elbow.

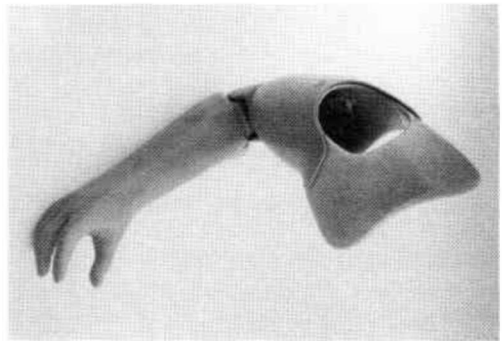


Fig. 2. The HMRC "mini" elbow in prosthesis.

from conventional prostheses. Because the available 62-type elbow would have been too large, a pair of electric elbows were made up from the chassis of Bock 6 ¾ size hands. The thumbs of the hands were mounted on the socket, the index and middle finger became the root of the forearms. The elbows were controlled by switches as described above. As the family insisted on hands, she was given two VASI 2–6 hands which were controlled with 3-state systems. Within a couple of days this girl, not yet 3-years-old, learned to control the elbows and hands and was feeding herself. However, there were maintenance problems with these make-shift elbows and they were replaced later with the first 3–8 "mini" elbows which had been designed by the HMRC mechanical engineering section (Figs. 1 and 2). Today, the girl is a very effective user of her arms showing a remarkable degree of skills at home and in school. She is, however, unique only because she was the youngest patient to be fitted with electric elbows. Equally good results have been observed in children, teenagers and adults with shoulder disarticulation, amelia or forequarter level amputations when using electric elbows and hands.

Prosthetics and Orthotics International, 1991, 15, 96–99

The stump capping procedure to prevent or treat terminal osseous overgrowth

J. PFEIL, E. MARQUARDT, T. HOLTZ, F. U. NIETHARD, E. SCHNEIDER, and C. CARSTENS

Orthopädischen Universitätsklinik, Heidelberg, Germany

Abstract

Terminal overgrowth in transverse deficiencies and in amputations, particularly of the humerus and tibia necessitate serial trimming procedures or re-amputation. Capping using an autogenous bone graft or a titanium and polyethylene endoprosthetic cap provides a satisfactory way of avoiding these re-amputations and allowing end-bearing.

Introduction

In transverse diaphyseal deficiencies as well as in acquired amputations in children bony overgrowth is a troublesome condition. The children have pain at the end of the stump, weight-bearing is impossible on the lower limb, and there is the risk of cutaneous perforation with secondary infection. The conventional practice in treating bony overgrowth has been the resection of the overlying bursa and shortening of the bony stump or in some cases re-amputation at a higher level. The child shown in Figure 1 has had a total of 6 re-amputations resulting in a short stump necessitating prostheses of the shoulder disarticulation type.

Stimulated by Swanson (1972) who developed a silicone rubber implant for capping an amputation stump, Marquardt (1976) developed his technique of an autogenous stump cap in 1974 and in 1985 he designed an endoprosthetic stump cap. The goal was to convert a diaphyseal deficiency or amputation into a stump resembling a disarticulation type.

All correspondence to be addressed to J. Pfeil, Orthopadische Universitatsklinik, 6900 Heidelberg, Schlierbacher Landstrasse 200A, Germany.

The procedure

A transplant source is not usually available in a case of a single limb deficiency, but in the quadrimembral deficient child various reconstructive procedures of other parts may make a transplant source readily available. For example in a PFFD the femural remnant can provide the necessary cartilaginous and bony cap. In traumatic amputations a distal epiphysis may be used. This is also possible in a planned amputation, for example in an osteosarcoma of

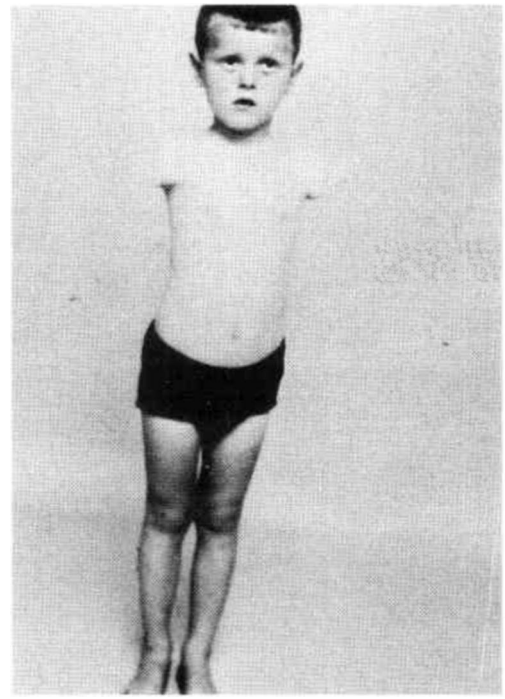


Fig. 1. A 12 year old boy, short humeral stumps afer 6 re-amputations due to terminal osseous overgrowth

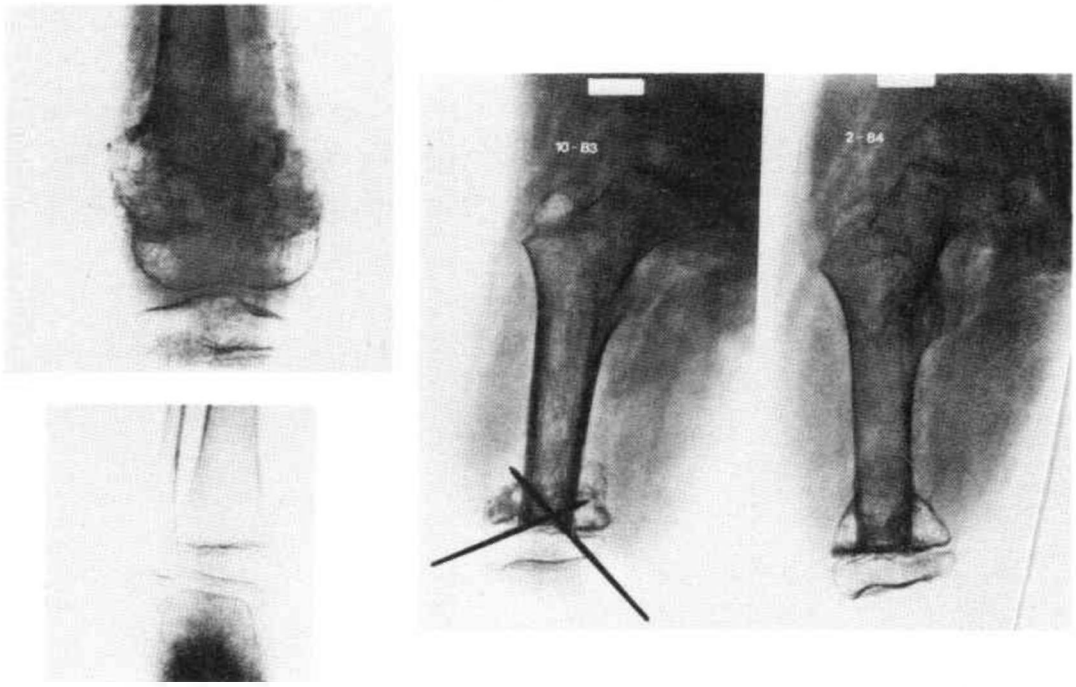


Fig. 2. Osteosarcoma in the distal femur in a 10 year old boy. The distal epiphysis of the tibia is used as an autogenous stump cap at the time of amputation.

the distal femur (Fig. 2). Alternatively a graft may be taken from the dorsal part of the iliac crest. The surgical procedure consists of the resection of the bony spike, then the transplant is fixed with a screw or K-wires. The periosteum is reattached to the transplant, the remaining place is then filled with cancellous bone (Fig. 3).

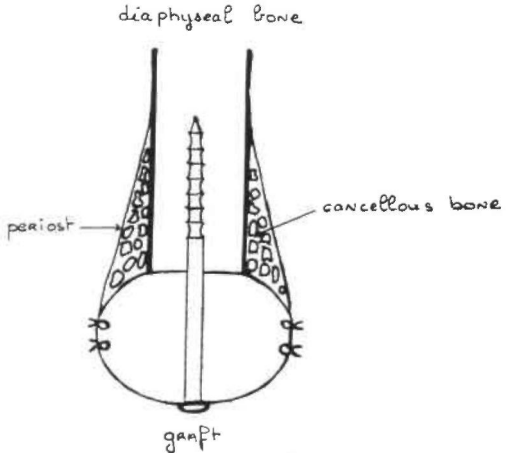


Fig. 3. The technique of the autogenous stump capping procedure.

A medial longitudinal incision is made a few centimetres proximal to the end of the stump so as to avoid scarring the end-bearing area. The bony spike is resected and the cap fixed with K-wires or with a cancellous screw. To give you an idea of this procedure Figure 4 illustrates the follow-up X-rays of a 4 year old girl with very short epiphyseal stumps in bilateral PFFD. In her case the cartilaginous bony caps were preserved from the femur and a stump capping procedure was performed on both arms at the age of 4 years. A good end-bearing capacity was achieved on both sides. Eight years after the operation the patient was able to bring her arms together in front of the body. Following the stump capping procedure the skin at the end of the stump became tight but there was no risk of bony perforation.

In this early case the incisions were made at the end of the stumps which is thought to be responsible for the narrowing of the soft tissues. When she was grown up a musculo-cutaneous flap was performed to give better soft tissue covering of the stumps. This procedure has, in our opinion, only cosmetic benefits, and does not bring any functional improvement.

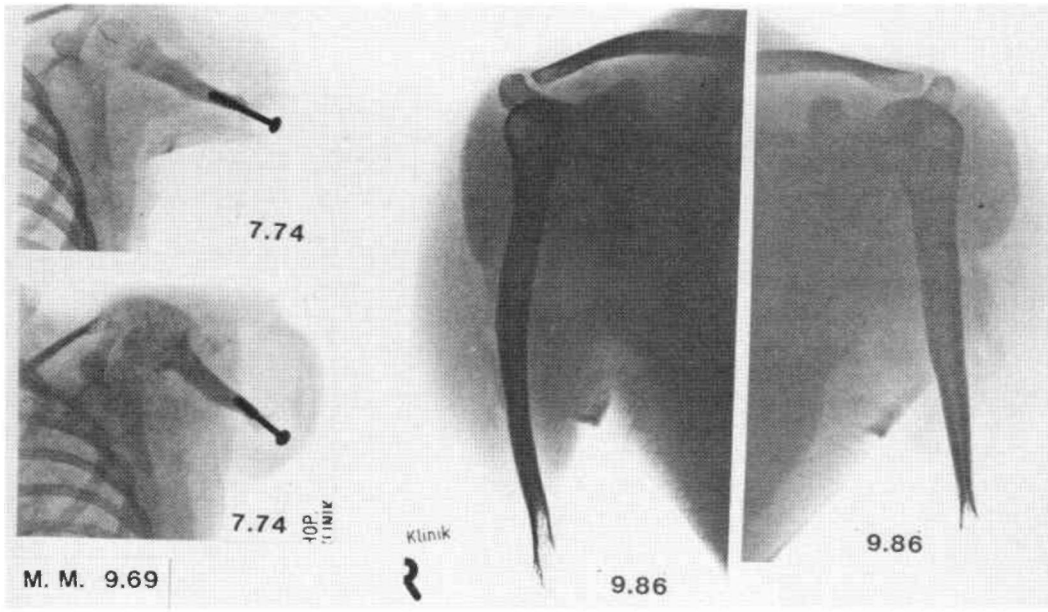


Fig. 4. The use of osseo-cartilaginous transplant from the femur to the humerus bilateral, in a quadrimembral deficient child. Notice the growth of the humerus after this procedure in the follow up films.

It is important to recognize that the success of this procedure depends not only on the surgical technique but also on a postoperative training programme. Approximately 3 months after the stump capping procedure a programme of end-bearing training is started. Initially weights of 2 or 3 kg are used and gradually increased until the patient is able to take at least 50% of body weight directly over the stump end. Immediately after the end-bearing training the therapist teaches the patient the technique of skin stretching distally over the stump to prevent contractures or tightening of the skin over the reconstructed end. The prescription of and training in the use of the prostheses is also part of the postoperative care.

The advantage of the autogenous stump capping is that no artificial material is needed, and it produces good long term results.

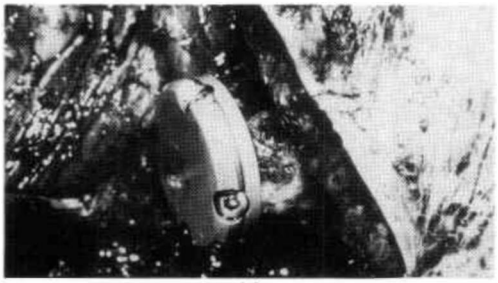
There are some disadvantages in this procedure. In some cases a graft is not available. At lease two incisions are necessary. Fixation metal has to be removed at a later stage. Sometimes consolidation takes a long time, especially in cases where a purely cartilaginous transplant is used.

In 1985 Marquardt developed in co-operation with Philipp Hannover, and the MECRON Company an endoprosthetic stump cap which is produced in different sizes. It consists of a titanium body which is fixed with at least three cortical screws to the bone. A polyethylene head covers the fixed titanium cap.

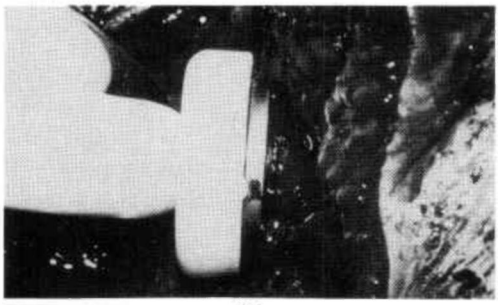
Figure 5 shows the application of the endoprosthetic stump cap. First musculo-periostal flaps are raised and the spike resected, before the titanium cap is fixed with cortical screws. A polyethylene cap is fixed with a self-locking system. At the side of the polyethylene cap there are holes which allow the fixation of the musculoperiostal flaps. In this particular case it was possible to close the muscles over the polyethylene cap. Full end-bearing capacity at the femoral stump could be achieved. This method is also applied in short humeral deficiencies or in short tibial stumps.

The advantages of the endoprosthetic stump cap are that no graft is needed and it is possible to achieve early weight-bearing. End-bearing can be started approximately 3 weeks postoperatively. A definitive prosthesis can already be used by the patient 6 weeks after this procedure.

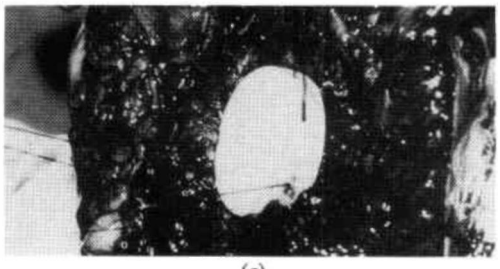
So far there is a lack of long term results. At present the number of cases treated in the University Clinic and a short postoperative follow-up prevents the provision of the complication rates of this procedure.



(a)



(b)



(c)

Fig. 5. Application of the endoprosthetic stump cap:
(a). fixation of the titanium body:
(b). fixation of the polyethylene cap with a self-locking system:
(c). attachment of the muscles to the polyethylene cap.

REFERENCES

MARQUARDT E. (1976). Plastische Operationen bei drohender konchendurchspiessung am kindlichen Oberarmstumpf. Eine Vorlanfige Mittelung. *Z. Orthop.*, **114**, 711–714.

SWANSON, A. B. (1972). Silicone-rubber implants to control the overgrowth phenomenon in the juvenile amputee. *ICIB*, **11 (9)**, 5–8.

Prosthetics and Orthotics International, 1991, 15, 100–103

The treatment of longitudinal radial deficiency

D. W. LAMB

Princess Margaret Rose Orthopaedic Hospital, Edinburgh, UK.

Abstract

Absence of the radius is the commonest of the longitudinal deficiencies but is itself quite an ususual congenital anomaly. No one surgeon is likely to encounter the condition more than once or twice in a career and there is a strong case for congenital upper limb conditions to be collected into special clinics.

The pathology and methods of surgical treatment are reviewed. It is a difficult condition to treat but there is no doubt that function and appearance can be improved (Pulvertaft, 1973). Pollicisation of the index finger should be considered and can often greatly improve the function.

Longitudinal deficiency

Radial Club Hand is the commonest of the major longitudinal deficiencies. It results from hypoplasia and complete or partial absence of the radius. Complete absence is the commonest and may be bilateral or unilateral. The condition was common in cases of thalidomide embryopathy when two-thirds of the radial deficiencies were bilateral. Its aetiology is otherwise unknown. It is usually sporadic but rarely may be associated with other congenital abnormalities: 1) Thrombocytopenia (the TAR syndrome). (It is the only time that absence of radius is associated with a near normal thumb); 2) Fanconi syndrome; 3) Atrial septal defect (the Holt-Oram syndrome). (This is the only condition where absence of the radius is hereditary); 4) VATER syndrome (vertebral defects, anal atersia, tracheo-oesophageal fistula, and radial and renal dysplasia).

The deformity is characterised by a radially deviated wrist and the forearm is short, only growing to between half and two-thirds of normal length (Fig. 1). The elbow usually lies extended at birth and there is a varying degree of reduced active and passive flexion at the elbow which often improves during the early years of life. Operation to correct the deformity should be avoided, particularly in bilateral cases, until the elbow flexes actively to at least 90 degrees.

The defects spread beyond the absence of the radius (Heikel, 1959) and stiffness of the elbow and there is stiffness and contracture in the metacarpophalangeal and proximal interphalangeal joints (Fig. 2). The scaphoid and trapezium often fail to develop and the first ray may be missing or represented by a simple "dangle thumb" (Pouce Flottant) (Fig. 3). This usually has a deficient metacarpal and no long tendons or thenar muscles. Growth potential is poor and reconstruction is not recommended. The radial forearm muscles are severely affected; the flexor pollicis longus is usually absent, the index profundus often absent or attenuated, though the flexor carpi radialis is usually present but the radial carpal extensors are poor or absent. The extensors to the index and middle fingers are usually hypoplastic and often have abnormal insertions limiting the metacarpophalangeal movement. Neuro-

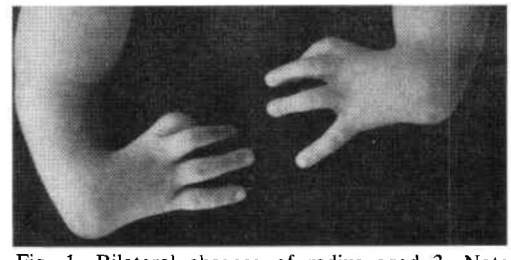


Fig. 1. Bilateral absence of radius aged 3. Note shortened forearm. The first ray of hand is absent and there is a four fingered hand with contracture of the radial two digits at the p.i.p. joints.

All correspondence to be addressed to Douglas W. Lamb, c/o Department of Orthopaedic Surgery, Princess Margaret Rose Orthopaedic Hospital, Fairmilehead, Edinburgh, UK.

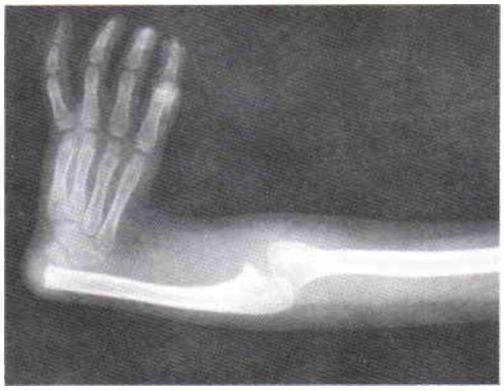


Fig. 2. Radiograph showing absent radius and severe club hand deformity in the untreated wrist. No orthosis had been used to maintain the wrist straight and the deformity was uncorrectable.

vascular anomalies are frequent and the terminal part of the radial nerve may be missing and the area of sensation supplied by that nerve taken over by an abnormal division of the median. The radial vessels are often absent and also the radial digital artery to the index, which has implications if pollicisation is planned,

Treatment

The deformity is usually correctable at birth and gentle stretching by mother and therapist is indicated. Corrective splintage is difficult but once possible should be used at night and the baby allowed to use the hands normally during the day. These measures should be continued until a decision is made regarding operation. The wrist deformity is so ugly and the functional disability in bilateral cases so great that operation should be considered. The optimal time is between 6 and 9 months of age especially if it is intended later to pollicise the index finger.

Numerous operations described over the past 100 years have been inevitably followed by relapse of the deformity (Lamb, 1977). The operation with the best long-term improvement has been centralisation of the carpus (Lamb, 1977; Lidge 1969) (Fig. 4). Recurrence is liable during growth and an important part of the operation is transferring a major deforming force, by the flexor carpi radialis, to the dorsoulnar aspect of the wrist. Splintage of the wrist in the corrected position post-operatively is important and should be maintained for a minimum of 6 months and then at night for several years during growth.

Buck-Gramcko (1985) positions the ulna on the radial side of the carpus, (radialisation) thus avoiding resection of carpal bones.

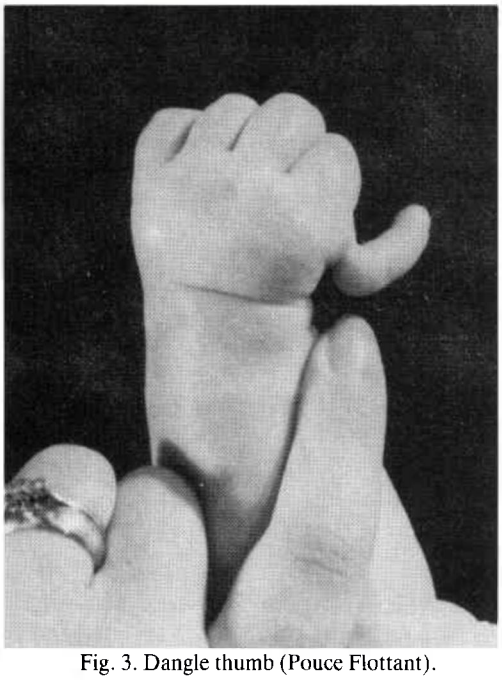


Fig. 3. Dangle thumb (Pouce Flottant).

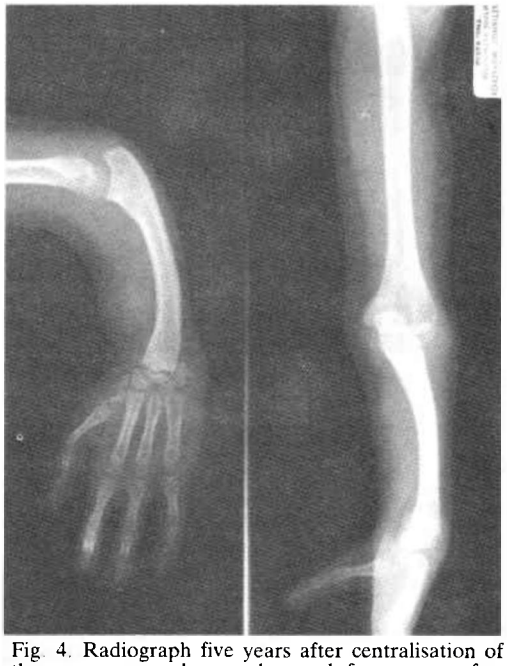


Fig. 4. Radiograph five years after centralisation of the carpus over lower ulna and four years after pollicisation of index finger.

Operation

The main steps are similar for both centralisation and radialisation. 1) An 'S' shaped incision is made from dorsum of hand to volar aspect of forearm. This gives good access to the carpus, the lower end of the ulna, the median nerve and the forearm muscles. 2) The extensor retinaculum is raised from radial to ulnar side and left attached to the ulnar side, and the extensor tendons exposed. 3) The median nerve is identified and any anomalous branch to the radial side. 4) The radial forearm muscles are exposed. The flexor carpi radialis can usually be identified but is sometimes joined with the radial carpal extensors. These muscles are removed from their insertion and will be transferred to the dorsi-ulnar aspect of the wrist later in the procedure. 5) An osteo-periosteal flap is raised from the dorsum of the carpus and the lower end of the ulna is dissected extra-periosteally without damage to the epiphyseal cartilage or its blood supply. Any fibrocartilaginous remnant of the radius (occasionally present in partial absence of radius) should be excised with all fibrous tissue preventing full passive repositioning of the hand over the ulna. 6) The next step depends on the nature of the procedure. If centralisation is intended the cartilaginous carpus is identified, sufficient of the central portions of this is removed to insert the lower ulna to a depth equal to its transverse diameter. This is in no way an arthrodesis. The intention is to preserve limited movement at this new ulno-carpal joint. The osteoperiosteal flap from the dorsum of the carpus is closed firmly and provides stability. It it is unstable or if any corrective osteotomy of the ulna is necessary, a Kirschner wire is inserted in a retrograde manner along the 3rd metacarpal shaft and then advanced across the centre of the lower end of the ulna and up the medullary cavity. This will keep the wrist correction and stabilise any ulnar osteotomy that was required. This wire can be left for several months but may extrude prematurely or break at wrist level with increasing activity. If radialisation is to be performed the ulna is positioned over the radial side of the carpus. A Kirschner wire is inserted obliquely through the second metacarpal and passed proximally along the shaft of the ulna. 7) The previously isolated radial muscles are transferred to the dorsi-ulnar aspect of the wrist and attached to the extensor carpi ulnaris. The retinaculum is repositioned and sutured in place.

Buck-Gramcko (1985) shortens the extensor carpi ulnaris by over-lapping and excises the redundant skin at the ulnar side. He reviewed 121 corrective procedures performed since 1969. Centralisation was carried out in the first 10 years and radialisation in the latter 10 years. Two-thirds had a subsequent pollicisation after the wrist operation. Results depend on the severity of the deformity, the condition of the muscles and function present in the fingers. He has little doubt that correction of the wrist is worthwhile, and that function is improved by pollicisation even though the quality of the index is seldom normal in this condition.

Lamb (1977) showed that children with bilateral deformity were unable to carry out all activities of daily living pre-operatively. Their overall function was significantly improved by correction of the wrist and pollicisation of the index finger.

Pollicisation (construction of a thumb)

A functional thumb is usually considered to contribute about 50 per cent of overall hand function. Attempts to reconstruct a functional thumb have therefore attracted much surgical endeavour (Littler, 1976). This is usually required for the following congenital conditions; 1) the four fingered hand; 2) the five fingered hand; 3) the "dangle thumb" with or without associated absence of the radius.

The technique evolved is based on transfer of the radial digit on its neurovascular bundle (Littler, 1953), shortening of the 2nd metacarpal and associated transposition of its interossei to reconstruct a new abductor and adductor of the thumb, and rotation of the retained 2nd metacarpal head so as to prevent an ugly hyperextension deformity of the thumb (Buck-Gramcko, 1971). Provided the structure and movement of the joints of the index finger and the quality of the intrinsic muscles of the 2nd ray are good the appearance and function of the index finger transposed in this way to the thumb position have given outstanding results.

Those surgeons who do not have the experience or expertise to obtain such results should refer children to a specialist unit.

Technique of operation

The operation in congenital absence is totally

different in concept and technique from operation for traumatic loss. Reconstruction of the whole thumb including the basal joint is required and is achieved by retaining the 2nd metacarpal head to form the new trapezium. Because of the wide range of movement at the metacarpophalangeal joint of children a hyperextension deformity is liable to develop at the base of the new thumb unless the metacarpal head is rotated into hyperextension so as to tighten the volar capsule.

Enough of the 2nd metacarpal shaft is removed so that the tip of the new thumb will reach the proximal interphalangeal joint of the long finger. If too much metacarpal shaft is preserved the thumb will look unnaturally long and be more a finger in the thumb position.

To obtain a good pinch between thumb and long finger the 2nd ray is rotated on its long axis about 150 degrees. The thumb should be abducted about 40 degrees.

If the intrinsic muscles of the 2nd ray are poor it will be necessary to reinforce these by an opposition transfer of the superficialis of the ring finger. This is seldom necessary with normal intrinsics which are stripped subperiosteally from the metacarpal shaft before its resection with careful preservation of their blood and nerve supply. These muscles are reinserted into the extensor aponeurosis over the original proximal interphalangeal joint which will now become the metacarpophalangeal joint. The first dorsal interosseus will become an abductor pollicis brevis and the first volar interosseus an adductor. The transposed radial intrinsic develops like a thenar eminence and in optimal cases the transposed index shows excellent mobility with full opposition and radial abduction (Fig. 5).

The optimal time for pollicisation is between 9 and 12 months of age. Buck-Gramcko (1971) has shown in 400 pollicisations for congenital cases that the end results are better the earlier the operation is performed. The transferred muscles hypertrophy, the bones of the index finger grow and broaden so that the proximal phalanx becomes like a first metacarpal and the child does not have to learn any new pattern of prehension.

For a good result the operation must be carried out meticulously and the surgeon have experience of working with small hands.

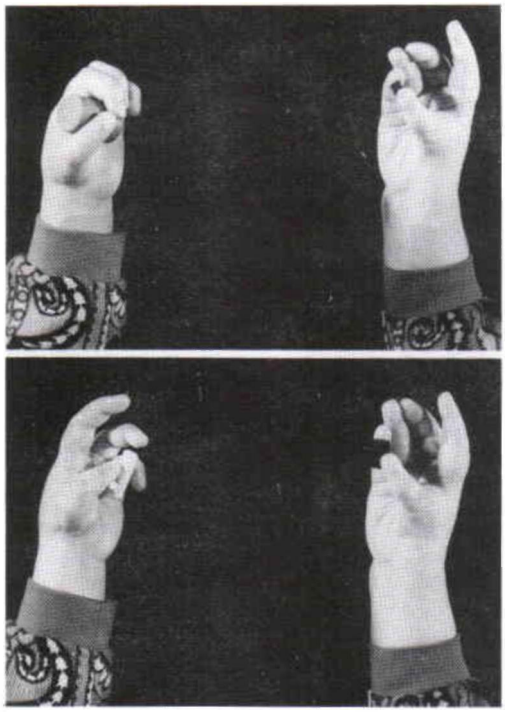


Fig. 5. Pollicisation of the left index finger at age 1 year. Excellent function with mobile thumb and good muscle control. Reconstruction right thumb with congenital absence thenar muscles.

REFERENCES

Buck-Gramcko, D. (1985). Radialization as new treatment for radial club hand. *J. Hand Surg.*, **10A**, 694–698.

Buck-Gramcko, D. (1971). Pollicization of the index finger. Methods and results in aplasia and hypoplasia of the thumb. *J. Bone Joint Surg.*, **53A**, 1605–1617.

Heikel, H. V. A. (1959). Aplasia and hypoplasia of the radius. Studies on 64 cases and on epiphyseal transplantation in rabbits with imitated defect. *Acta Orthop. Scand.*, **28**, (Suppl. 39).

Lamb, D. W. (1977). Radial club hand. *J. Bone Joint Surg.*, **59A**, 1–13.

Lidge, R. T. (1969). Congenital radial deficient club hand. *J. Bone Joint Surg.*, **51A**, 1041–1043.

Littler, J. W. (1953). The neurovascular pedicle method of digital transposition for reconstruction of the thumb. *Plast. Reconstr. Surg.* **12**, 303–319.

Littler, J. W. (1976). On making a thumb: one hundred years of surgical effort. *J. Hand Surg.* **1**, 35–51.

Pulvertaft, R. G. (1973). Twenty-five years of hand surgery, *J. bone Joint Surg.* **55B**, 32–55.

Prosthetics and Orthotics International, 1991, 15, 104–105

The treatment of longitudinal ulnar deficiency

R. J. LOVETT

Mary Free Bed Hospital and Rehabilitation Center, Grand Rapids, USA.

Abstract

Longitudinal ulnar deficiency, whose detailed anatomy is varied, is often accompanied by other abnormalities and appears as part of several syndromes. The history of its classification is reviewed, and the treatments, which have been offered, described.

Follow-up of a series shows that the function achieved is good and is optimised by aids, occupational therapy and some hand surgical procedures. The more elaborate surgical reconstructions are unlikely to be beneficial.

Introduction

Ulnar longitudinal deficiency is a post-axial abnormality of the upper limb in which the ulna is completely or partially absent. Not only is it the least common of the four major ray deficiencies, but it is also the most variable in its manifestations. Although it is usually sporadic in its occurrence, there have been occasional reports of genetic involvement. The reported incidence is 1:100,000 live births.

Classification

Since its first description over three hundred years ago, multiple and varied terminology have been used; at present, ulnar ray deficiency or ulnar hemimelia being the most popular. The correct terminology using the ISO/ISPO classification is longitudinal deficiency ulna, either partial or total.

Longitudinal ulnar deficiency is usually accompanied by shoulder, wrist, and hand abnormalities. The elbow may be in acute flexion, extension, or even present with a radio-humeral fusion. The shoulder is frequently unstable with scapular deficits (Figs. 1 and 2).

Most classification schemes have emphasised the anatomic and radiologic abnormalities

All correspondence to be addressed to Raymond J. Lovett, Area Child Amputee Center, Mary Free Bed Hospital and Rehabilitation Center, 235 Wealthy St., Grand Rapids, Michigan 49503 USA.

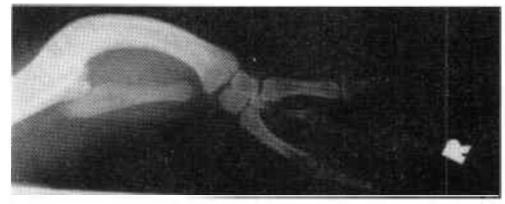


Fig. 1. A radiograph of longitudinal ulnar deficiency.

rather than the functional problems. Kummel (1895) classified this disorder by elbow anatomy. Specifically he characterised the radio-humeral joint as normal, fused, or dislocated. Ogden *et al.*, (1976) emphasised ulnar involvement using the terms hypoplasia, partial or total absence of the ulna. Swanson *et al.*, (1984) proposed a classification scheme based on both elbow and ulnar involvement. Rigault *et al.*, (1985) concentrated their classification scheme on hand function.

Classification is further complicated by the vast pot-pourri of additional abnormalities, including: other ray deficiencies, proximal femoral focal deficiencies, and several syndromes, especially the Cornelia de Lange and fibula-femur-ulna syndrome. In several studies, over half the patients had radio-ulnar synostosis. A third of the patients were bilateral, and there was a significant incidence of other limb involvement, both upper and lower. Eighty-nine percent (89%) of the patients has loss of at least one digit while 14% had a monodigital hand.

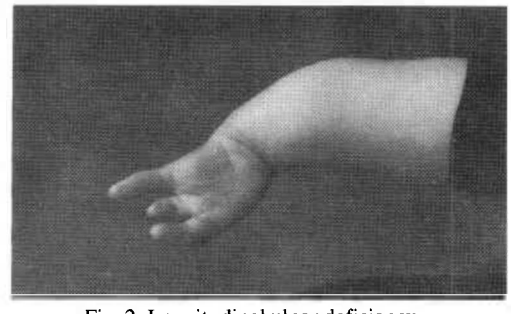


Fig. 2. Longitudinal ulnar deficiency.

Myths versus natural history

Most treatment has been aimed at improvement of function and there is general agreement that the standard hand and plastic surgical techniques, such as syndactyly release, webbed space deepening, and rotational osteotomies of the phalanges or metacarpals, when applied to this condition provide excellent functional improvement. However, some authors have recommended resection of the ulnar-distal radial cartilaginous anlage in an attempt to prevent shortening, bowing, and possible malrotation of the radius. More recent studies have shown that this procedure is rarely necessary and its use remains controversial. Another frequently advocated procedure is the creation of a one-bone forearm. This technique is applicable only when the proximal ulna is present and involves a radio-ulnar synostosis.

Since this procedure effectively eliminates pronation and supination, it has been shown in recent studies to compromise function rather than enhance it and is also no longer routinely recommended.

Recent quantitative functional studies have confirmed the older opinions that despite the anatomic and X-ray appearance, these patients function quite well without any surgical procedure other than those on the hand. Frantz and O'Rahilly (1971), in reviewing patients at the Area Child Amputee Center in Grand Rapids, Michigan, suggested prosthetic fitting with even possible elbow disarticulation in some cases. Further review of the literature has not revealed any groundswell of enthusiasm for prosthetic fitting and the amputation has been condemned. Recently, 61 patient charts at the Area Child Amputee Center, on which Swanson had previously done a demographic study, were reviewed to evaluate methods of treatment (Table 1). The majority of the patients had single limb involvement and no significant treatment was recommended. Other than hand or plastic surgery procedures, only seven surgical procedures were carried out. Two patients had humeral rotational osteotomies for cosmetic and functional improvement. Three patients had Z-plasty of the elbow without any significant evidence of improvement in motion, and two patients had elbow disarticulation. No cases of fibrocartilage remnant excision or one-bone forearm procedures were found. Humeral rotational osteotomies helped both cosmetically and functionally. The Z-plasties were a failure, but with today's improved microsurgical techniques should not be totally rejected. In the elbow disarticulation, one was performed for cosmesis and one was performed for function.

Table 1. Treatment of longitudinal ulnar deficiency.

Patients	61
Humeral rotational osteotomy	2
Elbow disarticulation	2
Elbow Z-plasty	3

In summary, other than the usual hand surgery techniques, these children are best treated by careful observation, the provision of adaptive aids, and emphasis on occupational therapy. Prostheses take away sensation and do not improve function. Rotational osteotomies may be of limited benefit.

REFERENCES

Birch-Jenson, A. (1949). Congenital deformities of the upper extremities. Odense, Denmark, Andelsbogtrykkerriet. p46–47.

Blair, W. F., Shurr, D. G., Buckwalter, J. A. (1983). Functional status in ulnar deficiency. *J. Pediatr. Orthop.*, **3,** 37–40.

Frantz, C., O'Rahilly, R. (1971). Ulnar hemimelia. *Artificial Limbs,* **15 (2),** 25–35.

Goller, D. C., (1698). Abortus humani monstros. *His. Anta. Misc. Acad. Nat. Curtos Norimb. Decuria,* **2,** 311.

Johnson, J., Omer, G. E. (1985). Congenital ulnar deficiency. Natural history and therapeutic implications. *Hand Clin.*, **1 (3).**

Kitano, K., Tada, K. (1985). One bone forearm procedure for partial defect of the ulna. *J. Pediatr. Orthop.*, **5,** 290–293.

Kummel, W. (1895). Die missbildugen der extremitaeten durch defekt Verwachsung und Ueberzahl. In: Biblioteca Medica, Section E, Vol. 3 — Cassell, T. G.: Fischer and Co.

Marcus, N., Omer, G. (1984). Carpal deviation in congenital ulnar deficiency. *J. Bone Joint Surg.*, **66A,** 1003–1007.

Ogden, J. A., Watson, H. K., Bohne, W. (1976). Ulnar dysmelia. *J. Bone Joint Surg.*, **58A,** 467–475.

Rigault P., Touzet, P., Padovani, J. P., Maroteaux, P., Finidort, G., Chaumien, D. P. (1985). Les hypoplasies et aplasies du cubitus chez l'enfant. Echonelies longitudinales internes du membre superieur. Etude de 38 cas chez 31 enfants. *Chirurgie,* **111,** 692–700.

Swanson, A., Tada, K., Yonenobu, K. (1984). Ulnar ray deficiency: its various manifestations. *J. Hand Surg.*, **9A,** 658–664.

Prosthetics and Orthotics International, 1991, 15, 106–111

The surgical treatment of congenital hand deficiency

P. J. SYKES,

Regional Plastic Surgery and Burns Unit, St. Lawrence Hospital, Chepstow, UK.

Abstract
This review describes the two types of congenital cleft hand deformity, stressing the important ways in which they differ and summarises the surgical techniques that may be used in their treatment.

Introduction

The two conditions are frequently grouped together under such titles as lobster-claw hand, ectrodactyly, oligodactyly and split hand. These terms are confusing, sometimes hurtful to parents and best discarded. Lange (1936) and Barsky (1964) have differentiated the typical from the atypical cleft hand and the two conditions are distinct, imposing a different surgical approach while sharing similar functional problems.

Central longitudinal failure of development: the typical cleft hand

Barsky (1964) credits Hartsinck (1770) with the first description of a case and Birch-Jensen (1949) claims that the first use of the term cleft hand was by Kummell (1895). The deformity (Fig. 1) has been described by Flatt (1977) as being "a functional triumph and a social disaster". It is rare and has been estimated at invovling 0.4 (Rogala, *et al.*, 1974) to 0.14 (Birch-Jensen, 1949) per 10,000 live births. Cases occur sporadically but the condition may be familial and is inherited as an autosomal dominant with mixed penetration (Graham and Badgley, 1954; David, 1974). Recessive inheritance may occur (Flatt, 1977). Both hands and feet are frequently involved and the cleft is 'V' shaped. Other anomalies sometimes affect the limb and the condition may be associated with abnormalities in various organs. Genetic counselling is particularly important for these patients (David, 1974). In the common form the middle finger is missing, with or without the metacarpal, and suppression of more digits in a radial direction occurs in other cases. Ring finger absence alone is rare and when the digits are so severely suppressed as to produce a single digital variety this is the little finger. Syndactyly frequently affects the digits bordering the cleft and where the thumb is involved this can severely affect function. Several metacarpal variations are described. This bone may be totally missing in the cleft or two metacarpals may seem to support one finger which then resembles severe compound syndactyly on radiographs. Alternatively a bifid metacarpal may support two digits. Several subclassifications have been proposed (Watari and Tsuge, 1979; Nutt and Flatt, 1981; Tada *et al.*, 1981). The cause of the defect is probably due to an abnormality in the apical ectoderm of the limb bud which produces a wedge shaped gap in the hand (Muller, 1937). The much quoted centripetal theory (Maisels, 1970) attempts to explain a progression of clefting from a simple central soft tissue gap to complete suppression of all digits.

Management

The first question is: do these patients need

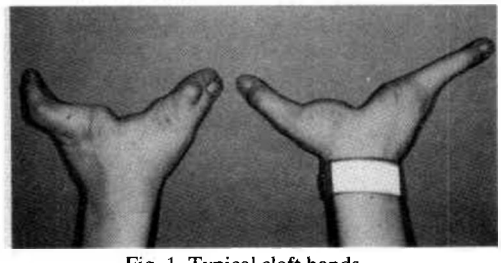


Fig. 1. Typical cleft hands

All correspondence to be addressed to P. J. Sykes F.R.C.S., St. Lawrence Hospital, Chepstow, Gwent NP6 5YX, UK.

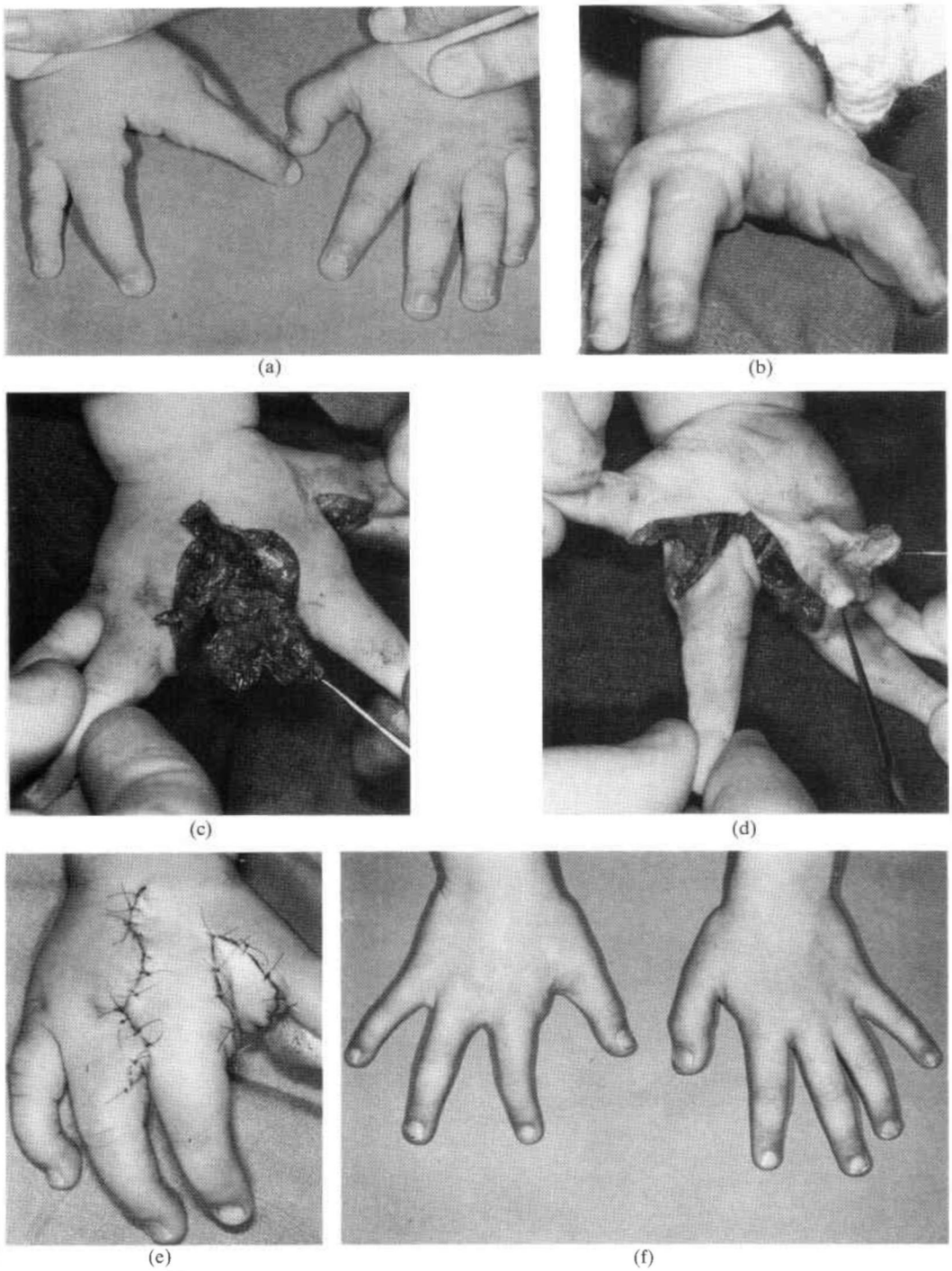


Fig. 2. Reconstruction of typical cleft hand by the Snow-Littler technique
(a) and (b) Pre-operative views
(c) and (d) Flap elevation
(e) Flap inset into thumb web and cleft closure
(f) Post-operative view

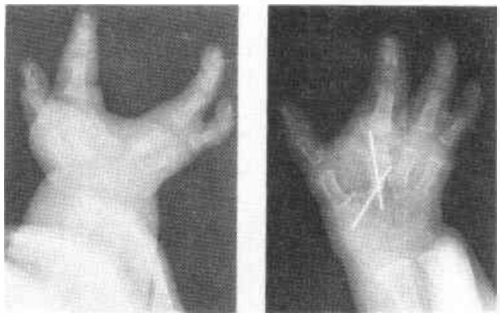


Fig. 3. Snow-Littler technique. Radiographs to show pre- and post-operative bony anatomy. The third metacarpal has been partially excised. The index metacarpal has been transected proximally, shifted ulnarwards and fixed to the base of the third metacarpal.

treatment? Cases which have not undergone surgery for a variety of reasons adapt and function extremely well often possessing excellent pinch and grasp despite deformaties which are very disfiguring. Experience has shown, however, that function can be improved by early surgery and this is usually performed between 1 to 2 years. Direct closure of the soft tissue cleft can be performed in minor cases and attempts should be made by using a local flap (Barsky, 1964) to reconstruct a web commissure. Recurrent finger separation by drifting of the border metacarpals following surgery should be prevented by the reconstruction of a transverse metacarpal ligament protected by temporary transmetacarpal Kirschner wiring. Free tendon grafts are frequently used for this technique and may be available locally as both the extensors and flexors of the missing finger are usually present in the ray and are fused over the end of the metacarpal remnant. If not, a palmaris longus graft can be used to hold the adjacent metacarpals together (Ueba, 1981). Frequently after excision of a redundant metacarpal and before closure of the cleft the collateral ligaments of the metacarpo-phalangeal joint need reconstructing using local tissue flaps including periosteum to prevent instability and deviation. In more severe cases where the residual digits are widely separated by a deep gap, spare skin from either side of the cleft should not be discarded. Local flaps fashioned from this tissue provide excellent skin for separation of the syndactyly which frequently affects the adjoining digits. This is a particularly valuable technique when the first web is involved with the thumb adducted and is the treatment of choice for the common situation where the middle finger is missing and the thumb partially fused to the side of the index. It can be used in combination with a shift of the index finger metacarpal in a ulnar direction with an osteosynthesis at the base of the middle finger metacarpal. The Snow and Littler procedure (1967) (Figs. 2 and 3) is an excellent technique which employs these two manoeuvres but other methods combining skin flaps from the cleft and a metacarpal shift have been described more recently (Miura and Komada, 1979; Ueba, 1981). For the less severe cases of central longitudinal deficiency the surgery is challenging, rewarding and gives excellent functional and aesthetic results (Fig. 4). Where suppression has been more severe the aims are less ambitious. When there are only two digits a combination of flap surgery, rotation osteotomy and tendon readjustment may restore a crude pincer grip. Where only 1 digit remains, treatment is more difficult and the results less satisfactory. Some of the methods discussed in the next section then become applicable to restore a simple prehensile function to the hand. These include microsurgical free toe transfer (May *et al.*, 1981).

Transverse failure of development distal to the carpus: the atypical cleft.

This condition is a different entity (Fig. 5). Treatment is generally more difficult and functional results less rewarding. The digital rays are missing at variable levels from the metacarpal distally. In less severe forms the thumb and little finger may be totally or partially preserved so that the child has a crude grasp and a useful pinch. The cleft of the hand is then 'U' shaped and the gap between the

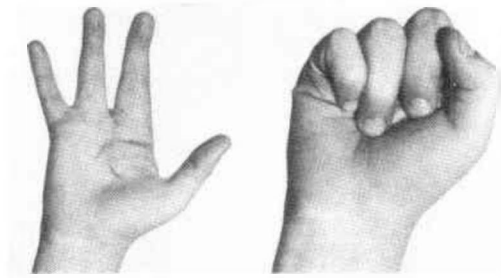


Fig. 4. Post operative function following Snow-Littler procedure for repair of typical cleft hand.

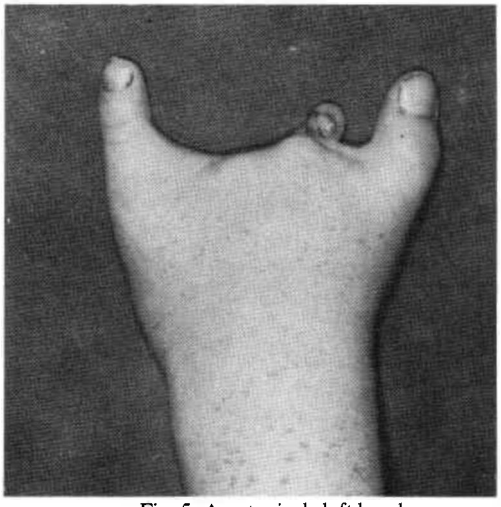
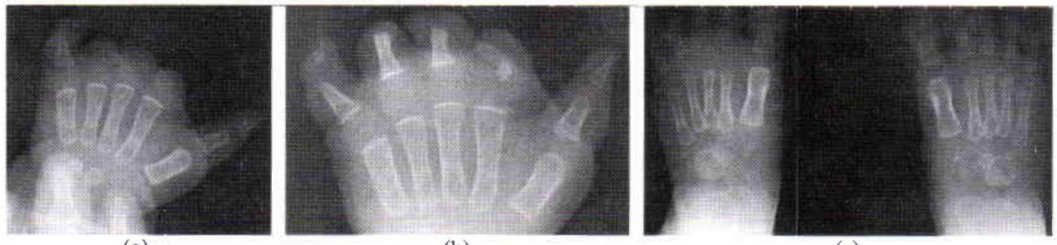


Fig. 5. An atypical cleft hand.

border digits is occupied by soft tissue nubbins with rudimentary nails representing finger remnants. This has led Buck-Gramcko (1971) to classify the deformity as a form of symbrachydactyly. The condition is sporadic and one hand is usually affected. Associated anomalies are less frequent. Unlike the typical cleft hand the monodactylous type of this deformity spares the thumb. The aetiology of the condition is probably a primary transverse failure of bone formation with subsequent disturbance of the soft tissue.

Management

Where bone and soft tissue are preserved distal to the carpus surgical reconstruction with the provision of sensate prehension has advantages over the use of an opposition device. Various options are available and each case must be considered separately after careful assessment of what is available and what is required. Simple excision of small nubbins of skin and deepening of the cleft between an adequate thumb and little finger may enhance function by a simple operation with low morbidity but when border digits are very short some lengthening operation must be undertaken. Free phalangeal transfer from the toes has had mixed success over the years (Carroll and Green, 1975; Goldberg and Watson, 1982) but the technique described by Buck-Gramcko and Pereira in 1990 in which a proximal toe phalanx is transplanted with its periosteum has been consistently successful (Fig. 6). These authors have shown that the phalanx survives and grows for a longer period if transplanted early and have been able to reconstruct a joint. The bone requires an adequate skin pocket for survival and when nubbins of tissue are large these should not be discarded too readily before considering this technique. Unstable digits with missing phalanges, in particular the thumb and little finger when a small terminal phalanx is unsupported on the metacarpal, can be lengthened and stabilised by a phalangeal transfer between the two bones of the digit. Alternatively a bone graft, which is less readily absorbed in this situation may be used. Other lengthening procedures are available but technically difficult. Rudimentary parts of digits including bone and soft tissue may be transposed on vascular pedicles to lengthen short adjacent fingers, and distraction lengthening of metacarpals has been reported (Smith and Gumley, 1985) but not gained widespread popularity. Complex reconstruction using a free bone graft covered by a skin flap to create an insensate opposition post for a



(a) (b) (c)

Fig. 6. An atypical cleft hand treated by phalangeal transfers from the toes.
(a) Pre-operative view
(b) Post-operative view
(c) Post-operative view of feet — proximal phalanges removed from left 2nd and 3rd toes. Middle phalanx removed from right 2nd toe.

solitary mobile digit is another option. This multi-staged procedure which is technically demanding does carry risks and has limited indications. A better alternative made possible by advances in microsurgery is the reconstruction of a digit using single stage toe transfer. The technique was first used following traumatic loss of the thumb. The great toe or the second toe is used but the latter has advantages in that it leaves a better donor site in the foot (O'Brien, *et al.*, 1978). The use of this method in reconstruction for congenital deformities is still in its infancy, but with improved microvascular technique survival is no longer the main problem (Gilbert, 1982; Lister *et al.*, 1983: Lister and Scheker, 1985). It seems likely that in carefully selected cases this technique will offer considerable benefit in the future. The operation is indicated for congenital amputations, especially of the thumb, when these occur in the "ring constriction syndrome". In these cases, nerves, vessels, tendons and bones have suffered an intrauterine amputation at the same level and are predictably available for reconstruction. The technique also has a place in the treatment of severe forms of transverse failure of development so that a useful prehensile pattern can be restored to the hand.

Conclusion

Typical and atypical cleft hand deformities occur with varying degrees of severity. Children learn to adapt from an early age and frequently develop remarkable function. If surgical treatment is used it should be carefully planned for each patient and performed early. Many techniques are described for improving hand function and, when the deformity is severe, these involve complex bone and soft tissue reconstruction.

REFERENCES

BARSKY A. J. (1964). Cleft hand: classification, incidence and treatment. *J. Bone Joint Surg.*, **46A**, 1707–1720.

BIRCH-JENSEN, A. (1949). Split-hand and atypical split-hand in congenital deformities of the upper extremities. Odense, Denmark, Andelsbog-trykkerriet. p 46–67.

BUCK-GRAMCKO, D. (1981). Allgemeines Wahloperationem. Handchirurgie. Band I. — Stuttgart: Thieme.

BUCK-GRAMCKO, D., PEREIRA, J. A. R. (1990). Proximal toe phalanx transplantation for bony stabilisation and lengthening of aplastic digits. *Ann. Chir. Main. Memb. Super.*, **9 (2)**, 107–118.

CARROLL, R. E., GREEN, D. P. (1975). Reconstruction of the hypoplastic digits using toe phalanges. *J. Bone Joint Surg.*, **57A**, 727.

DAVID, T. J. (1972). Dominant ectrodactyly and possible germinal mosaicism. *J. Med. Genet.*, **9**, 316–320.

DAVID, T. J. (1974). The differential diagnosis of the cleft hand and cleft foot malformations. *Hand*, **6**, 58–61.

FLATT, A. E. (1977). Cleft hand and central defects. In: The Care of Congenital Hand Anomalies. — St. Louis: C. V. Mosby. p265-285.

GILBERT, A. (1982). Toe transfers for congenital hand defects. *J. Hand Surg.*, **7**, 118–124.

GOLDBERG, N. H., WATSON H. K. (1982). Composite toe (phalanx and epiphysis) transfers in the reconstruction of the aphalangic hand. *J. Hand Surg.*, **7**, 454–459.

GRAHAM, J. B., BADGLEY, C. E. (1954). Split-hand with unusual complications. *Am. J. Hum. Genet.*, **7**, 44–50.

HARTSINCK, J. J. (1770). Beschryving van Guiana, of de wilde Kust in Zuid-America. — Amsterdam: Gerrit Tielenburg. Vol. 2, p811-812.

KUMMEL, W. (1895). Die Missbildungen der Extremitaeten durch Defekt, Verwachsung und Ueberzahl. In: Biblioteca Medica. Section E, Vol. 3. — Cassel, T. G.: Fischer and Co.

LANGE, M. (1936). Grundsatzliches uber die Beurteilung der Enstehung und Bewertung atypischer Hand-und Fussmissbildungen. *Verhandl. Deutschen Orthop. Gesellschaft*, **31**, 80–87.

LISTER, G. D., KALISMAN, M., TSAI, T. M. (1983). Reconstruction of the hand with free microneurovascular toe-to-hand transfer: experience with 54 toe transfers. *Plast. Reconstr. Surg.*, **71**, 372–386.

LISTER, G. D., SCHEKER, L. (1985). The role of microsurgery in the reconstruction of congenital deformities of the hand. *Hand Clin.*, **1**, 431–442.

MAISELS, D. O. (1970). Lobster-claw deformities of the hands and feet. *Br. J. Plast. Surg.*, **23**, 269–282.

MAY, J. W., SMITH, R. J., PEIMER, C. A. (1981). Toe-to-hand free tissue transfer for thumb construction with multiple digit aplasia. *Plast. Reconstr. Surg.*, **67**, 205–213.

MIURA, T., KOMADA, T. (1979). Simple method for reconstruction of the cleft hand with an adducted thumb. *Plast. Reconstr. Surg.*, **64**, 65–67.

MULLER, W. (1937). Die angeborenen Fehlbildugen der menschlichen Hand. — Leipzig: Thieme.

NUTT, J. N., FLATT, A. E. (1981). Congenital central hand defect. *J. Hand Surg.*, **6**, 48–60.

O'BRIEN, B. M., MACLEOD, A. M., SYKES, P. J., BROWNING, F. S. C., THRELFALL, G. N. (1978). Microvascular second toe transfer for digital reconstruction. *J. Hand Surg.*, **3**, 123–133.

ROGALA, E. J., WYNNE-DAVIES, R., LITTLEJOHN, A., GORMLEY, J. (1974). Congenital limb anomalies: frequency and aetiological factors. *J. Med. Genet.*, **11**, 221–233.

SMITH, R. J., GUMLEY, G. J. (1985). Metacarpal distraction lengthening. *Hand Clin.*, **1**, 417–429.

SNOW, J. W., LITTLER, J. W. (1967). Surgical treatment of cleft hand. In: Transactions of the Society of Plastic and Reconstructive Surgery: 4th Congress in Rome, Amsterdam, Exerpta Medica Foundation. p888–893.

TADA, K., YONENOBU, K., SWANSON, A. B. (1981). Congenital central ray deficiency in the hand — a survey of 59 cases and subclassification. *J. Hand Surg*, **6**, 434–441.

UEBA,Y. (1981). Plastic surgery of the cleft hand. *J. Hand Surg.*, **6**, 557–560.

WATARI, S., TSUGE, K. (1979). A classification of cleft hands, based on clinical findings: theory of developmental mechanisms. *Plast. Reconstr. Surg.*, **64**, 381–389.

Prosthetics and Orthotics International, 1991, 15, 112–116

The prosthetic treatment of lower limb deficiency

R. R. HIRONS, K. B. WILLIAMS, R. F. AMOR, and H. J. B. DAY

D.S.C. Withington Hospital, Manchester, UK.

Abstract

The prosthesis required for a child with a transverse deficiency whilst a simple version of that appropriate for the adult follows the same principles. The child with a longitudinal deficiency may require an extension, or ortho-prosthesis and this may be combined with surgical reconstruction. The principles, technique of measurement and fabrication methods using both traditional and modern composites are described.

Introduction

All cases of transverse deficiency save those whose level of loss is very distal require a prosthesis, whereas the prosthetic care of the child with a longitudinal deficiency may be combined with amputation or surgical reconstruction. Even after reconstruction of some of the major longitudinal deficiencies a prosthesis or orthosis may be necessary to achieve accurate length equalisation, improve the stability of joints and sometimes introduce some relief of weight bearing. However the device may be more efficient or better looking as a result of the surgery.

Whilst a lower limb prosthesis is not needed until the child is ready to stand and walk at perhaps 9–12 months, there is every need to plan the treatment and for the prosthetist to be involved at a very early stage, particularly when decisions about surgical reconstruction are being taken. It is only too common for the parent to have expectations which differ from those of the clinic team. The remark that leg lengthening will be done is heard and understood, but subsequent caveats that an orthosis may be required to brace an unstable joint are discarded by the parent who does not wish to hear this piece of information. The surgeon may recommend a procedure which allows the fitment of a prosthesis providing certain functional advantages, without realising that the relatives expect the prosthesis to look like a normal human leg.

Families need to be fully informed, understand and agree the treatment plan, and be made to feel integral members of the team. All must understand that the programme needs to be flexible, particularly as future joint development and even length discrepancy may be matters of conjecture in the early days. They must appreciate that their child's interest is not to acquire a good gait but to be able to "run" so that he can complete with his peers, as he must do if he is to achieve the overall objective of seeing himself as having the minimum possible handicap. Parents who do not agree to a proposal to amputate or reconstruct their child's leg must be allowed their view and should not be made to feel guilty.

Transverse deficiencies

The prosthetic treatment of those with transverse deficiencies and those whose longitudinal deficiencies have been converted by amputation can be discussed together. These latter will include ankle disarticulation for deficiency of the fibula and knee disarticulation for total tibial deficiency.

The prosthetic principles are the same as those used for older patients, though there would be no point in trying to incorporate, for example, a knee mechanism with sophisticated swing and stance phase controls in the prosthesis of a young child with a transverse thigh deficiency, indeed any knee articulation is unnecessary in the first few years.

Whilst it may seem desirable to use an ischial-gluteal rigid plastic socket for the above-knee case, the difficulty of taking a cast of the small child and the fact that he will grow out of the

All correspondence to be addressed to R. R. Hirons, C. A. Blatchford and Sons, D.S.C. Withington Hospital, Cavendish Road, Manchester M20 8LB, UK.

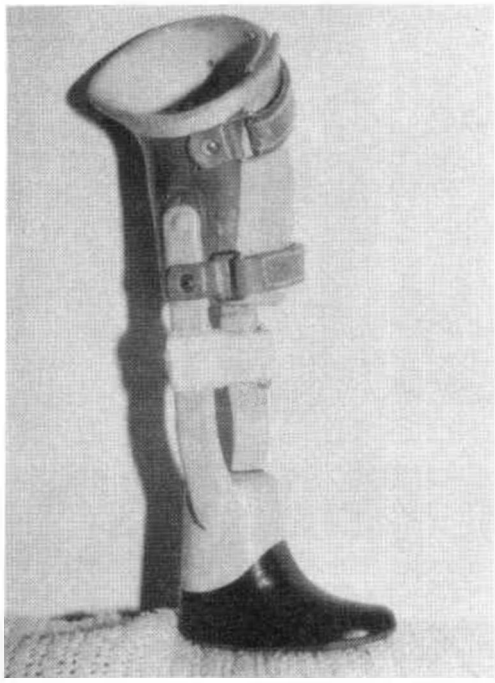


Fig. 1. Simple "first" prosthesis with leather socket.

socket very quickly often renders such a course impractical, and a leather socket which is capable both of adjustment and of being worn with diapers may be more satisfactory. Mounting this on aluminium side-bars above a foot provides a simple lightweight prosthesis (Fig. 1) which is easily lengthened. Sculpted foam between the side bars with an overall cover provides a satisfactory appearance.

A similar "long" prosthesis may be indicated as a first prosthesis for the child who has a deficiency below the knee. He can start active knee use when he is walking safely, though it may be considered wise to continue with some protection for the lower femoral epiphysis, by means of a thigh corset and side-bars and joints, and delay fitting a PTB prosthesis particularly in the child who has a short below-knee segment or is very active. In this case the lack of adjustability of the rigid socket and the child's growth in the early years may also pose logistic or financial difficulties. Conventional methods of construction, including the use of small size endoskeletal modular systems for all levels of loss, become appropriate when the child is older.

Longitudinal deficiencies

Those longitudinal deficiencies which have not been converted by amputation present with shortening as well as deformity, and it is the length inequality which must first be corrected if the child is to walk. Small length discrepencies, in the leg which is stable, can be corrected with a shoe raise, but if the discrepancy is greater or if there are joint defects, an extension prosthesis or ortho-prosthesis may be required.

Such a device will contain the child's leg and foot at a suitable height above a prosthetic foot (Fig. 2). The prescription requirements are set by the deformity, the ability to bear weight through the skeleton, and the power, range and stability of the joints. The amount of shortening and the alignment required will have a considerable influence on the angle of equinus which can be used and thus on the cosmetic appearance of the prosthesis. For example, in those cases with only moderate shortening the degree of equinus will be limited by the length of the child's foot, and the resulting appearance is poorer than that obtainable in a child with greater length inequality those own foot in equinus may be concealed in the prosthetic shin. In most cases the size and shape of the child's foot requires that the socket is opened to allow access, often by the use of a suitably reinforced leather bootee.

Day and Wright (1977) described the relationship between the clinical factors and

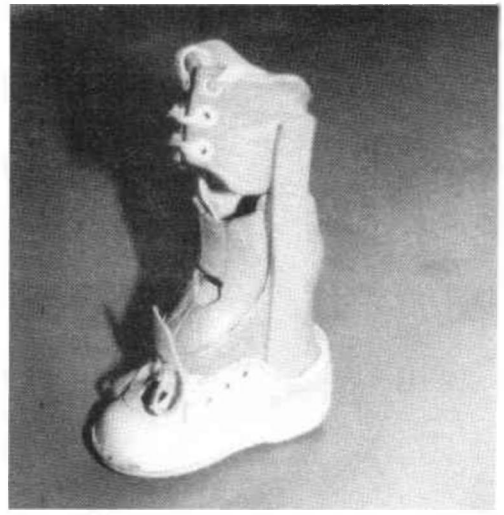


Fig. 2. Extension prosthesis with leather bootee and metal side-bars.

prosthetic prescription and demonstrated how this could be achieved using glass reinforced plastic prostheses to replace traditional leather and side-steel construction. Access to the rigid socket was achieved in one of two ways, either by a removeable access panel as in the Canadian Syme prosthesis (Foort, 1956), or by splitting the socket into anterior and posterior halves hinged together at the toe (Fig. 3). The socket is mounted at the appropriate height above a SACH foot or ankle mechanism, using a wood or rigid foam block after transferring out a child's size Berkeley alignment device.

In recent years the use of carbon fibre instead of glass as a reinforcement material in laminated polyester sockets has resulted in thinner, lighter and often cosmetically improved prostheses. This material has also been used to create a frame type socket which contains a leather bootee and replaces the simplest type of extension prosthesis consisting of a leather bootee reinforced by metal side-bars (Fig. 4).

Prostheses using carbon fibre in their construction are lighter and of improved appearance, but more frequent replacement may be required as adjustment of size and alignment to accommodate growth is limited.

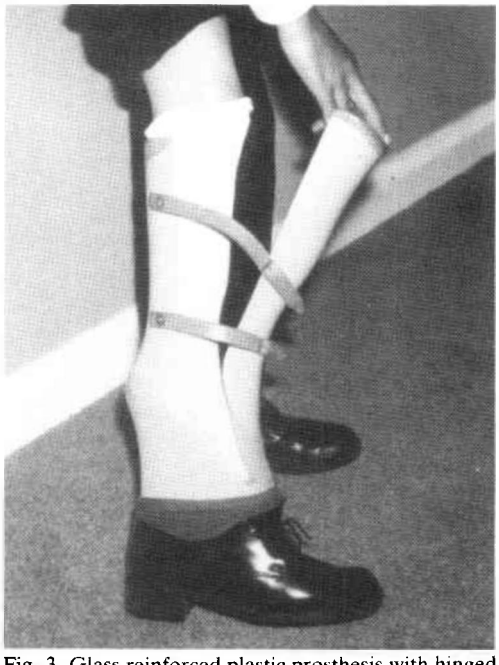


Fig. 3. Glass reinforced plastic prosthesis with hinged socket.

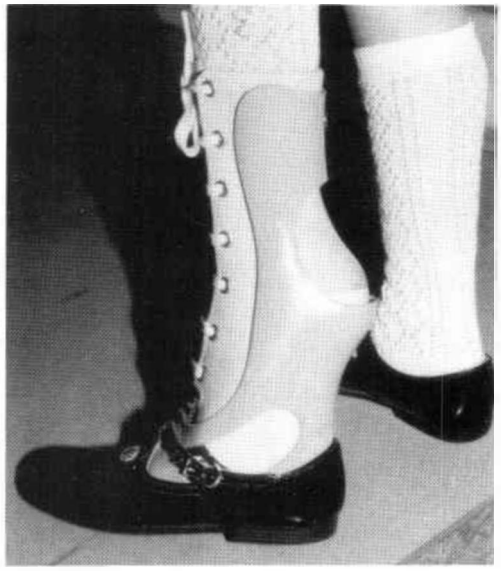


Fig. 4. CFRP frame with leather bootee.

These advances in fabrication techniques do not change the basic prosthetic principles which must be paramount, and the possibility of an enhanced appearance must not be allowed to compromise the optimum socket fit and alignment. For example, the flexion deformities of hip and knee accompanying proximal femoral deficiency must be accommodated and the child's centre of gravity maintained above the prosthetic foot without excessive lumbar lordosis (Fig. 5).

The desirability of this means of construction must be weighed against its cost and the fact that the required alignment is often unusual, sometimes bizarre, and not easy to determine without a trial fitting particularly when the infant is being fitted with his first prosthesis. At this stage traditional construction, using a leather "socket" mounted on aluminium side-bars has the advantage of simplicity and easy alteration.

It is important to explain to parents that adjustability in the early stages is essential and that a prosthesis with improved cosmesis can be produced once the child is walking and the optimum alignment has been established. Indeed the first device should not be referred to as a prosthesis or artificial limb, but as an aid to help their child to learn to walk.

The problems facing the prosthetist are formidable. The parents have expectations

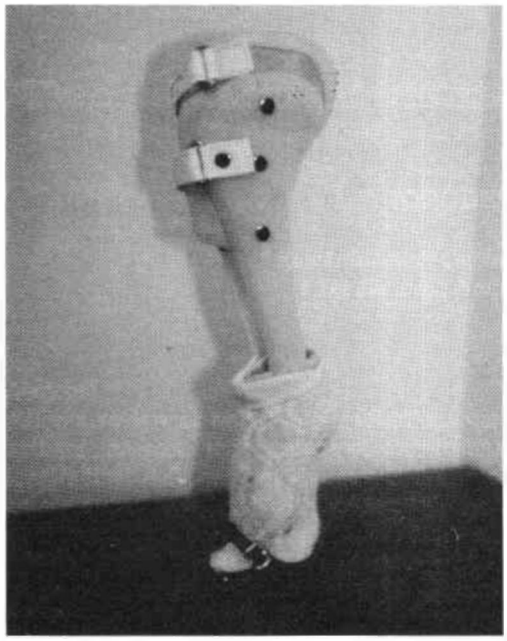


Fig. 5. Carbon fibre reinforced plastic frame with leather socket.

which are often higher than can be met, and he has to cast and measure a child who is only a few months old. He needs, perhaps, to take a full length cast encasing all the joints and to generate an ischial bearing fitting. This one limb is working in unison with three others and one small but perfectly developed mouth, all trying to keep the prosthetist as far away as possible.

A plaster impression is almost invariably needed to enable fabrication of any type of extension prosthesis, but the quality required of the cast depends on the materials to be used in the prosthesis. Thus a relatively poor cast may be adequate for the fabrication of the traditional type using leather and metal because these materials are easily adjustable. In contrast a high quality cast is mandatory if it is intended to produce a socket using reinforced laminated plastics or thermoplastics which fit very precisely and are not capable of effective alteration. The desirability of a high quality impression must be balanced against the likelihood of the prosthetist being able to achieve it. Dealing with complicated shapes is one thing, dealing with non-co-operation is another.

The prosthetist must have a plethora of techniques available for the young child, particularly on the first occasion. He has to decide whether to try and involve the child or divert its attention. The casting procedure can be segmental, starting distally, in order to immobilise each joint in turn and thus preventing destruction of the cast.

Liberal use of separating cream, apart from being fun, helps the removal of some thin casts in one piece. Deformation is minor and easily correctable at the time. Every effort is made to avoid the use of plaster shears or cutting tools for cast removal as young children remember unpleasant experiences and it will not be long before another visit will be needed.

Casts made in several sections to facilitate removal provide good quality replication but require a degree of co-operation which is unusual in a small child. However, these sessions become easier later when a relationship of trust develops between child and prosthetist and this is reflected in that between parents and prosthetist.

The manner in which the prosthesis is presented at the first trial fitting is vital. The

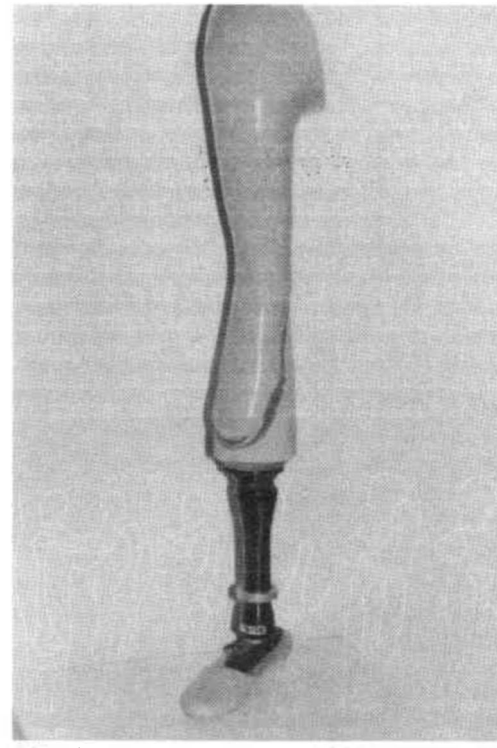


Fig. 6. Socket for child with PFFD mounted on endoskeletal components.

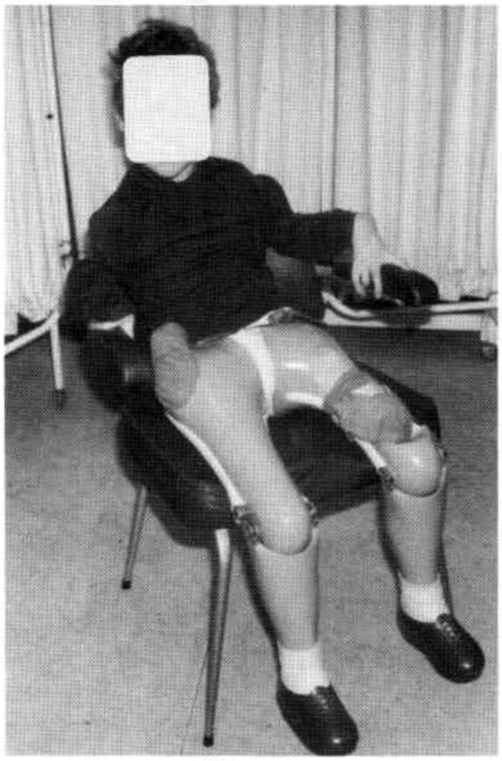


Fig. 7. Sitting position using Holmgren sockets.

parents' immediate reaction will dictate the mood for that session and influence future attitudes. Putting the child's own sock and shoe on the prosthesis before showing it and covering any necessary temporary junctions can be helpful. In short, no amount of technical expertise will make good any deficiency in the relationship between the prosthetist and family.

Fabrication using a wood block or rigid foam between the laminated socket and the foot is excellent for small children and those with little shortening, but older children and adults with considerable length descrepancy may benefit from the use of endoskeletal modular components distal to the socket (Fig. 6). This method of construction may provide a lighter prosthesis of improved function and appearance.

Children with bilateral gross longitudinal deficiencies whose hips are unformed or unstable pose considerable problems. Although experience shows that the majority will discard prostheses in adolescence, they should only do this on the basis of their experience. Simple flower pot types of sitting prostheses may be fitted quite early to enable the child to sit up and develop its skills. Later prostheses using swivel walkers or mechanisms similar to the parawalker or reciprocating gait orthosis may be used (Meadows *et al.*, 1990). Holmgren (1970) demonstrated a socket technique which provides good transfer of power and motion from the pelvis to the prosthesis which has proved very useful allowing some reciprocal as well as swivel gait, and enabling the wearer to sit, albeit in a semi-reclined posture, without the complications of hip locks (Fig. 7).

Summary

However fabricated, the fitting and alignment of the prosthesis must be based on sound biomechanical principles and an understanding of the effect of the deficiency on the anatomy, function and kinematics of the limb.

The use of modern materials and components from prosthetic systems has enabled the modern extension or ortho-prosthesis to be lighter, more efficient and better looking, though more costly than its predecessors.

Despite these advances the acceptance by the child and its family of the prosthesis depends as much on the relationship of trust built up between them and the professional members of the clinic team as on the type of fabrication and components used.

REFERENCES

Day H. J. B., Wright J. (1977). A system of extension prostheses. *Prosthet. Orthot. Int.* **1**, 8–12.

Foort J. (1956). Canadian type Syme prothesis. Berkeley, CA: University of California. Institute of Engineering Research. (Prosthetic Research Board Series II, issue 30).

Holmgren G. (1970). Prostheses for the lower extremities after severe bone malformation. *Prosthet. Int.* **3 (10)**, 20–29.

Meadows C. B., Stallard J., Wright D., Gordon L., Major R. E., Jones N. (1990) The Edinburgh-ORLAU prosthetic system to provide reciprocal locomotion in children and adults with complete transverse lower limb deficiency. *Prosthet. Orthot. Int.* **14**. 111–116.

Prosthetics and Orthotics International, 1991, 15, 117–126

The classification and treatment of proximal femoral deficiencies

I. P. TORODE* and R. GILLESPIE†

** Royal Melbourne Children's Hospital, Parkville, Australia*
† The Children's Hospital of Buffalo, Buffalo, USA

Abstract
After a review of previous classifications the authors suggest that cases of congenital longitudinal deficiency of the femur fall into one of two groups, each with specific features. The treatment alternatives and their indications are discussed in detail and a treatment plan suggested.

Introduction

The following paper attempts to present a balanced viewpoint of the management of congenital abnormalities of the femur based on the authors' experience and a review of the literature relevant to the topic as published in major orthopaedic publications.

The paper examines this topic under the headings of Classification, Clinical Problems, Surgical Procedures and Conclusions.

Classification

Standard radiographic techniques have been the basis of most of the classifications published to date, although the use of ultra-sound has recently been examined. One would expect that in the future the patients under discussion will be subjected to Magnetic Resonance (MR) scanning and no doubt further classifications will then arise.

At the present time however, the best known classifications in the English literature are those of Aitken (1968) and Amstutz and Wilson (1962). The original classification of Aitken contained four classes with the least severe being Class A and the most severe being Class D. The classification of Amstutz further subdivided these groups with Aitken's Class A being subdivided into Amstutz and Wilson Type I and II. However, from the original Amstutz and Wilson classification, further subtypes have arisen.

Hamanishi (1980) defined five types of deficiency (Fig. 1) and further subtyping resulted in the identification of ten groups of patients. More recently publications of Hillman *et al.* (1987) and Stihle *et al.* (1987) have provided further radiographic evaluation of patients with femoral abnormalities.

However, it is apparent in these papers that the radiographic appearance of children born with a congenital femoral abnormality is a continuum from the most minor variance from normal to the most severe deficiency of complete absence of the femur. Taken to its logical conclusion, the number of subtypes one can derive would simply depend on the number of patients that have been examined.

Some questions arise from the radiological classifications. Firstly, at what age should the classification be performed? Secondly, will this classification change with time? Thirdly, and perhaps most importantly, what relevance clinically are these numerous classifications to the patient and their ultimate limb function?

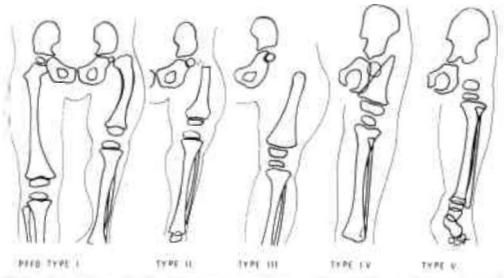


Fig. 1. Morphology, natural history and treatment of PFFD. Amstutz.

All correspondence to be addressed to I. P. Torode, Royal Melbourne Children's Hospital, Flemington Road, Parkville, 3052, Australia.

Table 1. Clinical signs of Group I patients

GROUP I
CLINICAL SIGNS

LEG LENGTH DISCREPANCY
20-30%

VALGUS KNEE

A-P LAXITY KNEE

FLEXION DEFORMITIES
NOT FIXED

FOOT MID TIBIAL LEVEL

Of relevance to the first question is the need to provide the parents of an infant born with a congenital abnormality with information that will be the foundation for their relationship with the doctor over the years to come. Indecision and/or incorrect information may well provide a permanent stumbling block in the doctor/patient relationship in the future and may well heighten the anxiety level of the parents who perceive this disability as a disaster for both themselves and their child.

In addressing the second question as to the change of classification with time, some information is gained by examining the expansion of the Amstutz classification with the passage of time. Hillman (1987), showed that with retrospective evaluation, only 17 out of 43 patients were correctly classified. This serves to illustrate that of itself radiographic evaluation may not be the foundation stone for the management of these patients.

The publication of Gillespie and Torode (1983) demonstrates that on clinical grounds patients with congenital abnormalities of the femur can be divided into two groups. The first of these groups contains those patients designated as having a congenitally short femur. Radiographically these groups could be classified as Aitken Class A or Amstutz Type I and possibly Type II. The clinical features of the Group I classification are shown in Table 1.

This group contrasts with the Group II patients who have more severe femoral deficiency and different clinical signs. Most of these patients would be classified as Aitken B, C or D groups at birth and show the unstable signs defined by Fixsen and Lloyd-Roberts (1974). The clinical signs of this group are summarised in Table 2.

This clinical classification becomes relevant when attempting to answer the third question posed. For those patients with congenital short femur, classified as Group I, there will be a possibility of performing surgical procedures to equalise limb lengths with the ultimate aim of avoiding the need for prosthetic fitting. This aim contrasts with the goals for those patients in Group II, true proximal femoral focal deficiency, for whom prosthetic fitting will be an inevitable part of the management. Exceptions to this are bilateral cases and where bilateral prosthetic fitting is to elevate the height of the wearer. In this situation the prostheses are often rejected.

Once the examining physician has ascertained into which of the broad clinical groups his patient falls it is necessary to examine the anatomical regions of the whole limb. The radiographic appearances of this femur and the presence of associated anomalies in the rest of the limb will dictate the variation in advice given. The radiographic appearances of the patients of Group I and II are summarised in Table 3.

Table 2. Clinical signs of Group II patients

GROUP II - PFFD
CLINICAL SIGNS

THIGH VERY SHORT

LEG LENGTH DISCREPANCY
35-50 %

FLEXED HIP AND KNEE
(FIXED)

ANKLE AT KNEE LEVEL

The limb

The foot and leg

The incidence of anomalies of the foot and leg said to be associated with congenital

Table 3. Radiological signs

GROUP I CONGENITAL SHORT FEMUR	GROUP II PROXIMAL FEMORAL FOCAL DEFICIENCY
FEMUR 40-60 % NORMAL	FEMUR VERY SHORT
NO DEFECT	DEFICIENCY ALWAYS NOTED
COXA VARA	HEAD AND NECK MAY BE ABSENT
LATERAL BOWING SHAFT	SHAFT MAY BE DEFICIENT
HYPOPLASTIC KNEE	HYPOPLASTIC KNEE

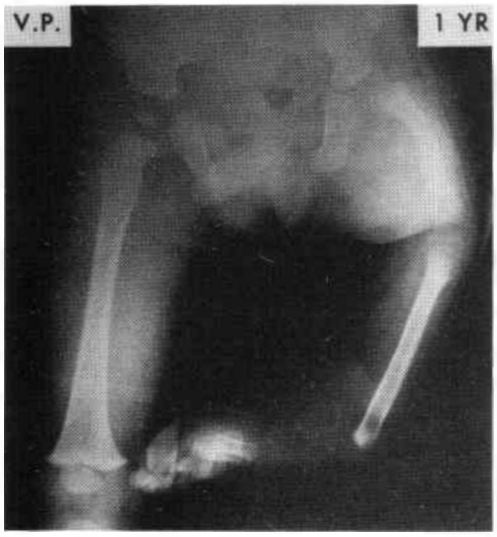


Fig. 2. Congenital short femur with associated fibular deficiency.

abnormalities of the femur varies from one report to another, but a reasonable assumption is that some abnormality will occur in approximately 50% of cases. The most common will be a variable degree of fibula deficiency (Fig. 2). This may range from a slightly short fibula in association with a valgus ankle, to the extreme case of a deficient foot with absent rays and a grossly unstable and displaced ankle joint. At either end of this spectrum, the desirability of keeping this foot will be obvious, but in the mid-range of deformities, other factors will be considered in deciding whether or not the foot is to be ablated.

Knee

Clinically the knee in these children will be held in flexion, but the flexion deformity of the Group I patients will become less with time, whereas the flexion deformity of the Group II patients will usually be permanent. This flexion deformity will tend to disguise the valgus attitude of the knee, often associated with a hypo-plastic lateral femoral condyle (Fig. 3). The knee in these children will also be unstable in an antero-posterior direction (Gillespie and Torode, 1983) due to the hypoplasia of the cruciate mechanism which has been documented by numerous authors, (Torode and Gillespie, 1983[1]; Johansson and Aparisi, 1983). Instability of the knee will become

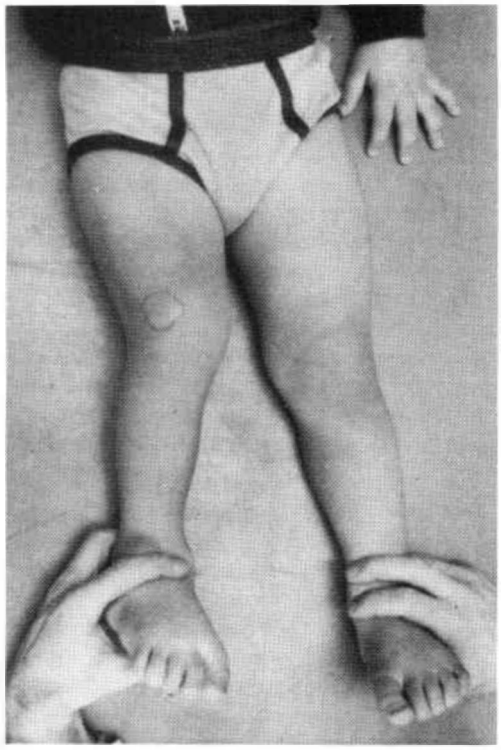


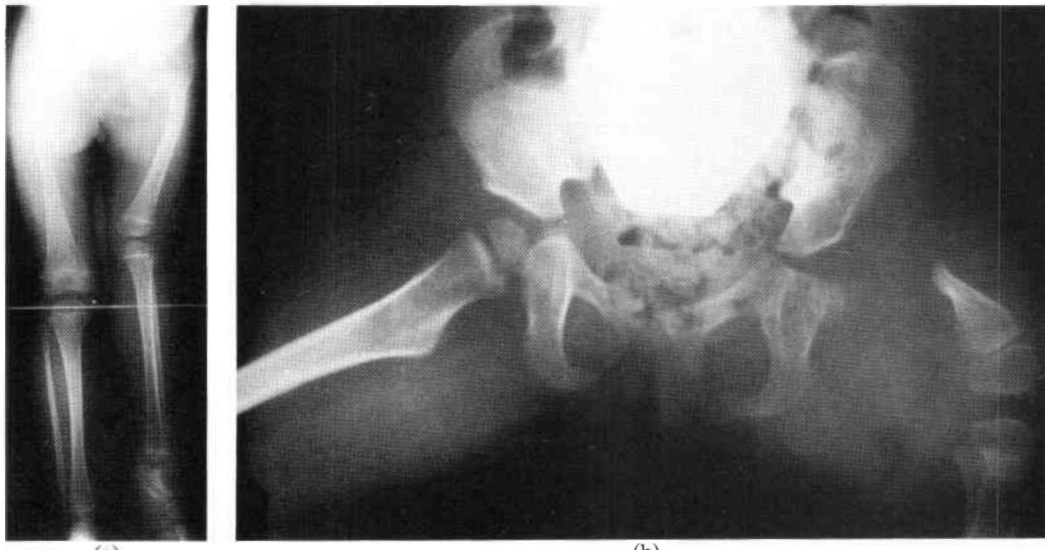
Fig. 3. Patient with right congenital short femur and valgus knee and external rotation deformity of the femur.

important in discussions surrounding lengthening procedures of the limb.

Thigh and hip

The varying degrees of deficiency of the proximal femur will be noted in simple radiographs. This may range from the case with a slightly short femoral neck, through those with a varus femoral neck, or an actual deficiency of the proximal femur distal to an intact femoral head and acetabulum, to a situation of gross deficiency of the proximal femur, femoral head and acetabulum (Fig. 4).

The radiographic appearance may vary with time and as further ossification occurs it will become apparent that the radiographic deficiency may have been a cartilagenous anlage which will go onto full ossification, albeit often in varus, and occasionally with an extreme shepherd's crook deformity. On other occasions this cartilagenous anlage may be absent or fail to ossify resulting in a definite pseudarthrosis in the proximal femur.



(a) (b)

Fig. 4 a) Left congenital short femur (Group I patient).
b) Severe right femoral deficiency (Group II patient).

Computer Tomography (CT) scans and X-ray examinations of the proximal femur also indicate a deficiency of the soft tissues surrounding the hip, and in particular a deficiency of the abductor musculature. One would expect that an MR scan of the hip would help to further delineate the soft tissue deficiencies.

The overall picture of the hip joint will play an important part in deciding whether bony reconstruction of the proximal femur is advisable and further knowledge of the soft tissue deficiency will assist the surgeon in understanding the limitations of the bony reconstructions as pertain to the final function of the hip joint.

Changes of the limb with time and growth

The authors believe it is possible on the basis of clinical signs to ascertain in most cases into which of the two clinical groups the patient should be placed, i.e. either a congenital short femur, or true proximal femoral focal deficiency. Despite the variations with age however, the radiographic appearance of the Group I femur in infancy will show the bulbous stable appearance as described by Fixsen and Lloyd-Roberts (1974), and with time further ossification of the proximal femur will proceed with varying degrees of coxa vara. The varus deformity however remains static throughout growth unlike true congenital coxa vara.

In Group II patients the actual deficiency of the bony elements will become apparent as ossification takes place. Nonetheless there will be a constant feature of a fixed flexion deformity of the proximal femur, although there may be an extension deformity through a pseudarthrosis in the subtrochanteric region in the femur (Fig. 5).

An important component of the parents' understanding of this clinical problem is the knowledge that the overall limb length deficiency will be relatively constant throughout growth. This predictability of

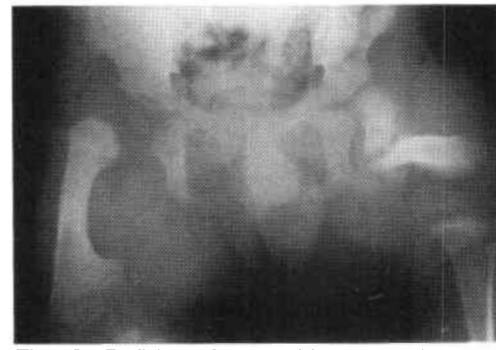


Fig. 5. Deficient femur with pseudarthrosis in extension in subtrochanteric region of the femur (Group II patient).

growth pattern has been well documented by Hamanishi (1980), Hillman (1987), Gillespie and Torode (1983). It is therefore usually possible to give the families an estimate of the expected limb length discrepancy that the child will have as he or she becomes more mature.

Using this knowledge the families can be advised at an early age of the various prosthetic appliances that can be utilised to assist the developing child in ambulation. However, as regards surgical procedures, particularly ablative procedures, the authors believe that these should be carried out earlier rather than later in the child's life. Clearly some surgical procedures are time dependent and the parents need to be made aware of the factors that the physician is considering in his decision making process for each particular child.

Surgical procedures

It is possible to look at the surgical procedures required in relation to the needs of each of the Groups I and II patients. There will, occasionally, be children who despite a deficiency of the proximal femur may be treated as Group I patients because of their particular anatomy, and there are others who have an intact femur, i.e. Group I, but whose femoral length deficiency is severe and beyond the limit of lengthening procedures. These therefore will be managed as Group II patients. It should be recognised that these are the exceptions rather than the rule and most can be managed utilising the procedures described below.

Surgical procedures appropriate to Group I patients

Syme's amputation

In the presence of an associated foot deficiency, with or without a fibular deficiency, it is usually readily apparent which children should undergo Syme's amputation and in which the foot should be retained. The greater the degree of the associated fibula deficiency and/or tibial deficiency the greater the likelihood of need for foot ablation.

The benefit of foot ablation is that there is a greater tolerance in the limb length equalisation procedures as a prosthesis can make up some of the deficiency in length and any small difference in the height of the knee axes above the ground is of no great consequence. This procedure should be carried out at about the time the child is attempting to stand and walk so that using a prosthesis becomes second nature early in life.

Proximal femoral osteotomy

Osteotomy of the proximal femur is a most useful procedure in those children who have an associated coxa vara. This procedure adds length to the limb by increasing the neck shaft angle and may also stimulate growth of the femur. Care must be taken not to over-do the valgus correction of the proximal femur, particularly if there is any degree of acetabular dysplasia. This procedure can be carried out in the first few years of life as a "once off" procedure utilising simple fixation techniques.

Distal femoral osteotomy

In children in whom there is significant valgus deformity of the knee, the authors recommend that this be corrected by an open wedge osteotomy, which can be held with cross K-wires and the ipsilateral fibula can be used as a bone graft to stabilise the opening osteotomy. This procedure will also slightly increase the length of the femur both by the opening osteotomy and the realignment of the limb and also by growth stimulation of the distal femur (Fig. 6).

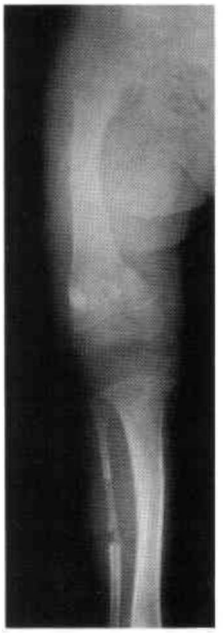


Fig. 6. Radiograph of Group I patient following opening osteotomy of distal femur and fibular graft.

Whilst it is recognised that the valgus attitude of the distal femur may be corrected at the time of femoral lengthening, the authors recommend for two reasons realignment of the limb as a whole prior to performing a femoral lengthening procedure. Firstly, the advantages of the osteotomy can be obtained at an earlier age than would be appropriate for a lengthening procedure. Secondly, it is much simpler to lengthen a straight segment than a deformed bone.

Innominate osteotomy

Careful assessment of the acetabulum must be made before performing any surgery on the proximal femur. Any residual dysplasia of the acetabulum can be corrected by a Salter innominate osteotomy, which must be performed before any lengthening procedures on the limb. Some additional gains in length can be obtained by using the Millis and Hall (1979) modification of the Salter innominate osteotomy.

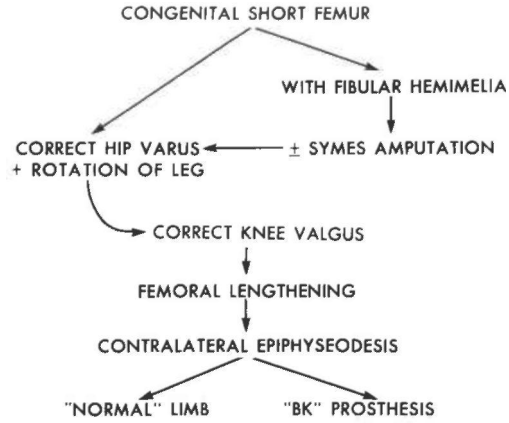
Limb length equalisation procedures

Utilising the knowledge that the proportions of the limb will remain constant throughout growth, one can estimate the expected discrepancy at maturity. The growth of congenitally deficient limbs can be monitored and graphed through the early years of childhood. By around 8 years of age, it will be evident as to whether the discrepancy can be made good by a single lengthening procedure with a contralateral epiphysiodesis, or whether it may be necessary to do two lengthening procedures.

Previously the authors have advocated that femoral lengthening be restricted to an increase of 20% of the original femoral length because of the risk of producing either dislocation or subluxation of the hip and flexion deformities or posterior subluxation of the unstable knee. With better understanding of the biology of bone as applied to bone lengthening and with the use of small wire external fixators, such as the Ilazarov or Monticelli Spinelli frames, the horizon has expanded and the complications reduced. Nonetheless it remains a procedure fraught with difficulties for the patient, parents and surgeon.

Epiphysiodesis may be used judiciously to reduce the femoral length and/or tibial length of the normal limb. The last few centimetres of femoral lengthening are often difficult to obtain and by reducing the ultimate length of the longer limb it is possible to lessen the complications and heartache. Modifications of the Phemister technique often leave ugly scars around the knee of the normal limb and this can be significantly improved upon by the use of the drill technique as described by Canale (1986). A flow diagram of the suggested plan of management for the Group I patients is shown in Table 4.

Table 4. Treatment plan for Group I patients



Surgical procedures in relation to Group II patients (PFFD)

This group of patients will always need prostheses if they are to achieve optimum function. The exception to this rule may be for these with bilateral deficiencies whose prostheses, used to elevate the patient to a more normal height, often prove unwieldy and are frequently rejected.

Syme's amputation

As discussed previously when the foot is deficient with respect to its lateral rays or tarsal bones, or if the ankle joint is grossly dysplastic in association with a severe degree of leg deficiency a Syme's amputation should be performed in the first few years of life (Fig. 7).

Where the foot is reasonably intact and the ankle joint functional, the families should be made aware of the functional benefits of the tibial rotationplasty and the part that procedure might play in the future management of the child. In such a situation the decision regarding a Syme's amputation must be delayed.

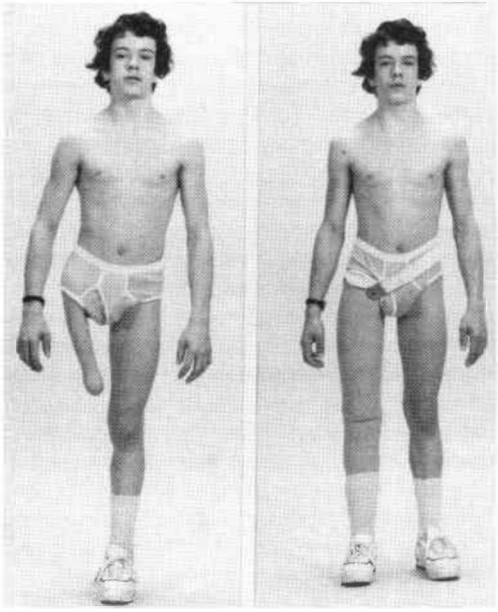


Fig. 7. Group II patient — following conversion to AK fitting by knee fusion and foot disarticulation.

Knee fusion

In cases where the families elect to have a Syme's amputation performed and the patient treated prosthetically as an above-knee amputee, the ablation of the knee joint adds considerably to the ease of prosthetic fitting and of prosthetic wearing. Figure 8 illustrates the difficulties that patients face when wearing an extension or above-knee prosthesis associated with a mobile hip and knee and with flexion deformities of both those joints. It is possible to perform a knee fusion early and preserve either the distal femoral or the proximal tibial growth plate, thereby ensuring further growth of the limb and the maintenance of a long stump. This procedure is performed by excising the appropriate segment of the knee joint and if it is intended to preserve the growth plate, the corresponding cartilaginous portion of the epiphysis is removed down to the cancellous bone of the ossific nucleus. Fixation is obtained by a smooth intramedullary rod passing along the length of the femur into the tibia. Compression devices have not been needed and fusion has been readily attained. This procedure can be performed in association with the Syme's amputation without undue difficulty.

Sometimes the knee fusion has been performed in association with a repair of proximal femoral deficiencies using the intermedullary rod of the knee fusion to provide fixation across the proximal femur. Knee fusion can also be performed as part of the Van Nes rotationplasty (Fig. 9). Patients in whom knee fusion is not to be performed are those for whom a femoro-iliac arthrodesis may be appropriate.

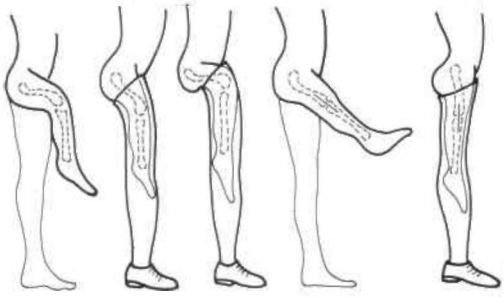


Fig. 8. Diagrammatic presentation of problem that is created by a flexed knee with prosthetic fitting of a short thigh segment.

Proximal femoral surgery

The enormous variation in radiographic appearance of the femora of Group II patients suggest an equally vast variety of surgical procedures that might be devised. In reality,

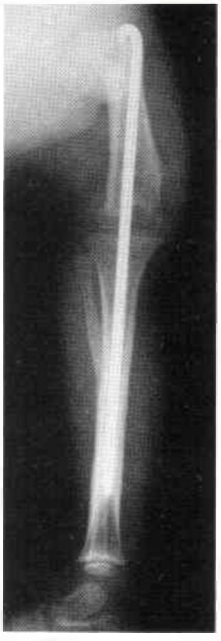


Fig. 9. Radiograph of Group II patient following rotationplasty. Note open proximal tibial growth plate.

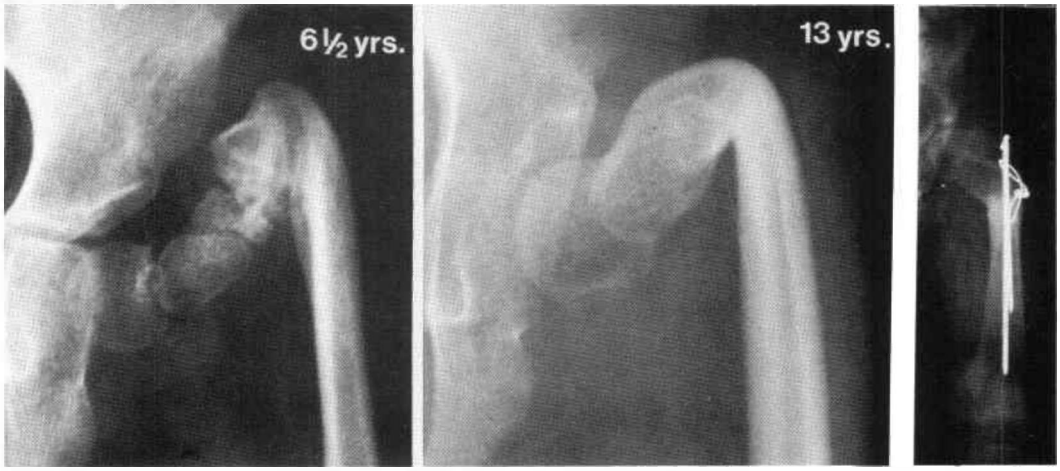


Fig. 10. Radiographs of Group II patient showing "shepherd's crook" deformity corrected by reconstructive osteotomy.

however, there are relatively few which are useful for the proximal femur of these patients.

The authors have not found exploration of the hip joint itself of great value. Exploration to confirm the appearance of the cartilaginous anlage will do little to further the function of these patients.

However, if there is instability or deformity due to a subtrochanteric pseudarthrosis of the femur, instrumentation and bone grafting to provide stability and a solid segment will be beneficial to the patient. Correction and internal fixation of gross varus or shepherd's crook deformity of the proximal femur may also improve the structural integrity of the bone and prevent recurrent stress fractures in the proximal segment of the femur (Fig. 10). A Trendelenburg gait due to the soft tissue deficiency, particularly of the gluteal and abductor musculature will often remain despite correcting the anatomical alignment of the proximal femur.

Tibial rotationplasty

The operative details of this procedure are described by Torode and Gillespie (1983[2]). However, it is worthwhile considering a few specific points. It is possible to perform the rotationplasty utilising the knee joint to attain either all or most of the rotation necessary. It is also possible to perform this operation at an early age and maintain one or both of the growth plates adjacent to the knee joint to ensure an appropriate length of the new "thigh segment" (Fig. 9). By rotating through the knee joint rather than through the tibia. the function of the anterior and posterior compartments of the leg are preserved and the tendency to de-rotate is markedly diminished.

The post-operative course is simplified and prosthetic fitting is more readily accommodated if the rotationplasty is performed early in life. There is no doubt that the end result of this surgery produces an unusual appearance, but there is little to differentiate these patients from the rest of the population when the prosthesis is being worn (Fig. 11). Furthermore the

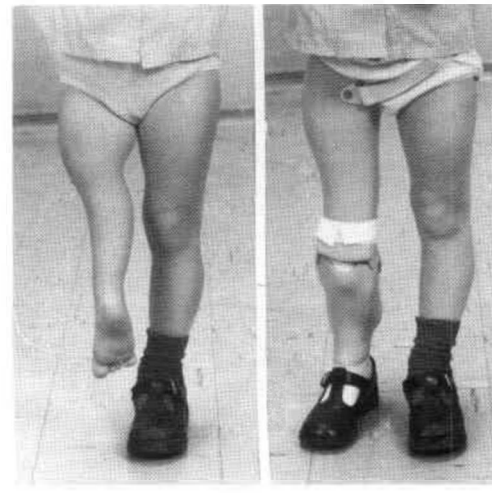


Fig. 11. Group II (PFFD) female patient following rotationplasty and knee fusion. Note long thigh segment to allow for growth discrepancy between legs.

functional advantages are quite significant and must be given due regard in the decision making before surgery is performed.

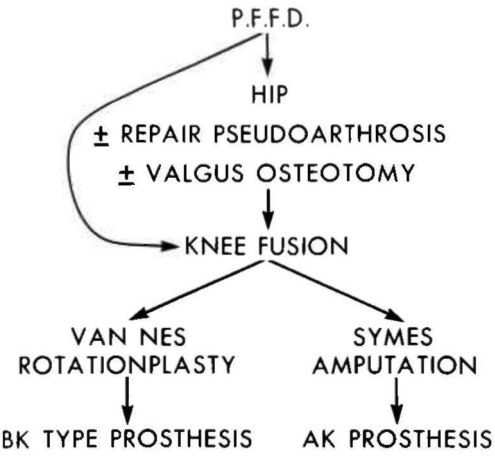
Fixsen (1983) presents the advantages and disadvantages of this procedure. Many of the problems he mentions have been addressed by the modifications of the technique as described by Gillespie and Torode (1983). Fixsen also raises the questions of psychological problems associated with the child having a foot at the end of the leg in a reversed position. However, it is difficult to differentiate between the embarrassment and psychologicial sequelae of the foot position after rotationplasty and the same sequelae of simply having a foot, often deformed, at the level of the contralateral knee, or if Syme's procedure has been performed, having no foot at all. The authors have interviewed all the children and young adults described in the rotationplasty group of their paper (Gillespie and Torode, 1983) and despite being offered foot removal, no patient has asked for this to be done to alleviate cosmetic or psychological concerns.

Iliofemoral fusion

Some of the patients with a true PFFD, exhibit a grossly deficient acetabulum and an absent femoral head resulting in a major problem of instability at the hip. Some may develop a spindle shaped end to the proximal aspect of the femoral segment which can ride upwards and be palpable just under the skin adjacent to the wing of the ilium. It may be appropriate to perform an iliofemoral fusion on such patients.

There are two important technical points in this procedure. Firstly, the femoral segment must be flexed approximately 90 degrees to its normal anatomical position, so that when the femur is arthrodesed to the ilium, the knee joint in extension will allow sitting and in flexion will allow standing. Secondly, the femoral segment must be reasonably short. If the femoral segment is too long when arthrodesed to the ilium, the weight bearing line of the limb will be at a considerable distance anterior to the coronal plane of the trunk and contralateral limb. However, many patients for whom this procedure appeared attractive have a knee flexion contracture and thus will not be able to extend the joint to allow a sitting position.

Table 5. Treatment plan for Group II patients



A more complete discussion on this procedure is given by Steel *et al.* (1987).

A summary of the surgical procedures described previously is shown in the treatment plan in Table 5. Despite the great variation in radiographic appearance, it should be noted that the number of useful surgical options is relatively limited and a decision needs to be made early in life to aim for below-knee function by use of a tibial rotationplasty or above-knee function by foot ablation and knee arthrodesis.

Summary

This paper has presented an overview of the management of congenital deficiencies of the femur. The two broad treatment patterns have been outlined, i.e. for children with a moderately severe deficiency, the congenital short femur group, and secondly children with a very severe deficiency with either a miniature intact femur, or a true proximal femoral focal deficiency.

Ideally, a child with a congenital short femur will be managed by re-alignment of the femur as a whole, followed by femoral lengthening with contralateral epiphysiodeses where appropriate. A Syme's amputation is reserved for those children in whom the foot is not a reasonably stable and functional component of the limb.

The child with a severe femoral deficiency and a significant foot deformity will be managed by a Syme's amputation and knee fusion and an above-knee prosthetic fitting. If it

is deemed appropriate, the child with a reasonably good foot may be treated by rotationplasty and knee fusion. The rotation being obtained through the former knee joint. The patient is then fitted with a rotationplasty prosthesis. For a very few patients with a grossly deficient femur and an inadequate hip joint, it may be advisable to perform an ilio-femoral fusion with the femur fused to the ilium in a flexed position. The child is then fitted with an above-knee type of prosthesis, with the former knee joint acting as the child's hip joint. It is not necessary to elucidate all the variations of femoral surgery that can be devised in these children, but where appropriate reconstruction of the proximal femur to overcome gross varus or to gain stability through a pseudarthrosis may be necessary.

REFERENCES

AITKEN, G. T. (1969). Proximal femoral focal deficiency — definition, classification and management. In: Proximal femoral focal deficiency — a congenital anomaly: a Symposium held Washington, D.C., June 13 1968. — Washington, D.C.: *National Academy of Sciences.* 1–22.

AMSTUTZ, H.C., WILSON, P. D. (1962). Dysgenesis of the proximal femur (coxa vara) and its surgical management. *J. Bone Joint Surg.*, **44A**, 1–24.

BOCHMANN, D. (1980). Prosthetic devices for the management of proximal femoral focal deficiency. *Ortho-Pros. (Can.)*, 4–29.

BORGGREVE, J. (1930). Kniegelenkseratx durch das in der Beinlangsachse um 180 gedrehte Fusgelenk. *Arch. Orthop. Chir.*, **28**, 175–178.

CANALE, S. T., RUSSELL, T. A., HOLCOMB, R. L. (1986). Percutaneous epiphysiodesis: experimental study and preliminary and clinical results. *J. Pediatr. Orthop.*, **6**, 150–156.

FIXSEN, J. A., LLOYD-ROBERTS, G. A. (1974). The natural history and early treatment of proximal femoral dysplasia. *J. Bone Joint Surg.*, **56B**, 86–95.

FIXSEN, J. A. (1983). Rotationplasty. *J. Bone Joint Surg.*, **65B**, 529–530.

GILLESPIE, R., TORODE, I. P. (1983). Classification and management of congenital abnormalities of the femur. *J. Bone Joint Surg.*, **65B**, 557–568.

HAMANISHI, C. (1980). Congenital short femur: clinical, genetic and epidemiological comparison of the naturally occurring condition with that caused by Thalidomide. *J. Bone Joint Surg.*, **62B**, 307–20.

HILLMAN, J. S., MESGARZADEH, M., REVESZ, G., BONAKDARPOUR, A. CLANCY, M., BETZ, R. R. (1987). Proximal femoral focal deficiency: radiologic analysis of 49 cases. *Radiol.*, **165**, 769–773.

JOHANSSON, E., APARISI, T. (1983). Missing cruciate ligament and congenital short femur. *J. Bone Joint Surg.*, **65A**, 1109–1115.

KING, R. E. (1968). Some concepts of proximal femoral focal deficiency In: Proximal femoral focal deficiency — a congenital anomaly: a symposium held in Washington, D.C., June 13 1968. — Washington, D.C.: *National Academy of Sciences.* 23.

KOSTUIK, J. P., GILLESPIE, R., HALL, J. E., HUBBARD S. (1975). Van Nes rotational osteotomy for treatment of proximal femoral focal deficiency and congenital short femur. *J. Bone Joint Surg.*, 1975, **57A**, 1039–46.

LANGE, D. R., SCHOENECKER, P. L., BAKER, C. L. (1978). Proximal femoral focal deficiency: treatment and classification in forty-two cases. *Clin. Orthop.*, **135**, 15–25.

MILLIS, M. B., HALL, J. E. (1979). Transiliac lengthening of the lower extremity: a modified innominate osteotomy for the treatment of postural imbalance. *J. Bone Joint Surg.*, **61A**, 1182–1194.

STEEL, H. H., LIN, P. S., BETZ, R. R., KLAMCHIA, C. M. (1987). Ilio-femoral fusion for proximal femoral focal deficiency. *J. Bone Joint Surg.*, **69A**, 837–843.

TORODE, I. P., GILLESPIE R. (1983[1]). Anteroposterior instability of the knee: a sign of congenital limb deficiency. *J. Pediatr. Orthop.*, **3**, 467–470.

TORODE, I. P., GILLESPIE, R. (1983[2]). Rotationplasty of the lower limb for congenital defects of the femur. *J. Bone Joint Surg.*, **65B**, 569–573.

VAN NES, C. P. (1950). Rotation-plasty for congenital defects of the femur: making use of the ankle for the shortened limb to control the knee joint of the prosthesis. *J. Bone Joint Surg.*, **32B**, 12–16.

WAGNER, H. (1971). Operative Beinverlangurung. *Chirurg.*, **42**, 260–266.

Prosthetics and Orthotics International, 1991, 15, 127–130

The surgical treatment of partial tibial deficiency and ankle diastasis

W. BLAUTH and P. HIPPE

Orthopaedic Department, Kiel University, Germany.

Abstract

Partial tibial absence and congenital ankle diastasis are described together with a surgical regime for their reconstruction.

Introduction

Operations performed on the foot and the lower leg in cases of tibial malformations require a comprehensive and detailed knowledge of the morphology of these most complex and variable deformities. The major morphological details are therefore described first.

The clinical appearance is, above all, characterised by a severe foot deformity, i.e. an equinovarus foot, as well as shortening and instability of the leg, along with a dysplasia of the knee joint (Fig. 1).

Partial tibial aplasia

The distal tibial epiphysis, metaphysis and the adjacent diaphysary section are missing. The diaphysis ends in the middle, sometimes in the lower third, of a thin and often pointed stump which may in addition also show a slight axial deviation. Sometimes, the skin in this region is scarred along with pitting, and the condition may be associated with polydactyly.

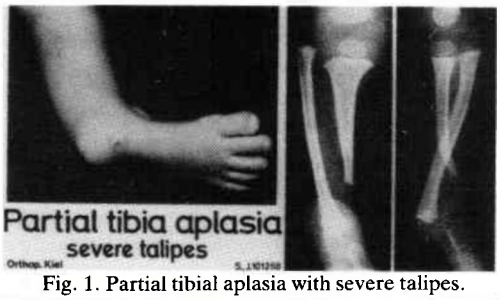


Fig. 1. Partial tibial aplasia with severe talipes.

All correspondence to be addressed to Dr. P. Hippe, Orthopaedische Universitatsklinik, Michaelisstrasse 1, 2300 Keil 1, Germany.

Lower leg diastasis

In lower leg diastasis, the tibial end is hypoplastic and does not articulate with the talus. It is even possible that the two lower leg bones override (Fig. 2). Parts of the tibia may be missing (Fig. 3), as already specified for the partial aplasia.

Interestingly enough, in these kinds of deformities the distal end of the tibia, provided with a soft tissue covering, my protrude from the residual lower leg, and there are similarities to the formation of clefts in hands and feet.

Foot

In both deformities the foot shows a distinct varus position. In some cases the sole points to the face. The talus is "suspended" at the distal fibular end in a tight capsulo-ligamentous interconnection. Feet in extreme equinus are predominantly seen in diastases of the lower leg. Tarsal synostoses, oligodactyly and even monodactyly are also found.

Editors Note:

The treatment of total tibial deficiency, first described by Otto in 1841, is either knee disarticulation or reconstruction. The former course is simple, requiring only one surgical procedure and allows the child to undergo straightforward prosthetic rehabilitation.

Reconstruction which seems an attractive option may require multiple surgical procedures and seldom obviates the need for an orthoprosthesis. It consists of transferring the upper end of the fibula to the intercondylar notch of the femur, and correcting the equinovarus deformity of the ankle by centralising the fibula into the talus. Furthermore it may be necessary to ablate the foot, so that the child becomes a below-knee amputee whose knee joint needs external support.

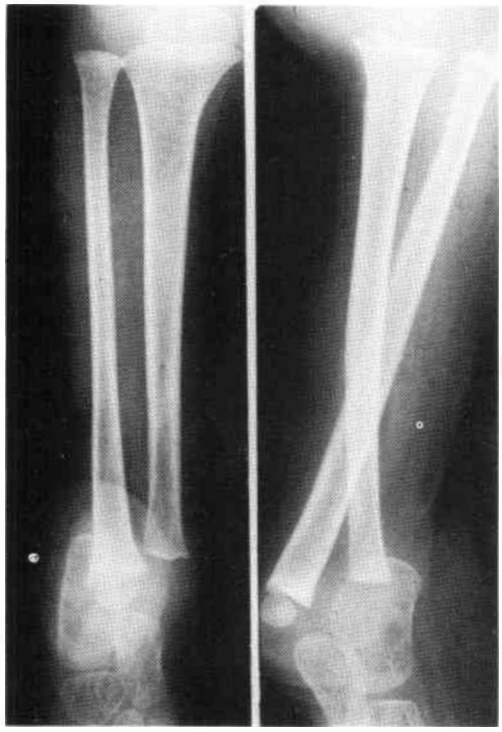


Fig. 2. Tibial hypoplasia with overriding bones in the lower leg.

Knee joint

An impaired knee joint development is evident with dysplasia of the articular components and patella, raised position of the fibular head and instability of the ligaments.

Indication for surgical treatment

Surgical treatment is indicated for all forms of tibial hypoplasia and partial aplasia, when it will provide consistently improved function of the deformed limb, allowing at the same time easier and better orthotic fitting.

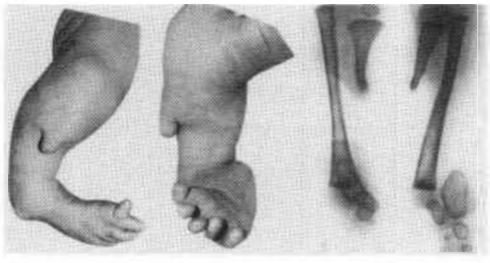


Fig. 3. Partial tibial aplasia.
Note: Distal end of the tibia contained in "skin bag", severe equinus foot deformity.

Surgery has two primary objectives:
- the correction of the foot deformity,
- replacement of missing tibial sections by the fibula.

Only after a stable and solid interconnection of foot and knee joint has been obtained is it possible to consider lengthening procedures. The authors usually proceed first to correct the deformed foot.

Surgical technique

In the early 60's the authors started to develop the so called foot realignment, with the hind foot being placed below the distal fibular end. The talus is then resected, with the fibula being placed in a recess in the calcaneus. The fibular epiphysis has to be prepared very carefully in order to preserve to the greatest possible extent the vascular supply of the growth line.

The foot is held in its position with Kirschner wires and a plaster cast. Immobilisation is required at least 8–10 weeks, although it is possible to remove the wire after 3–4 weeks.

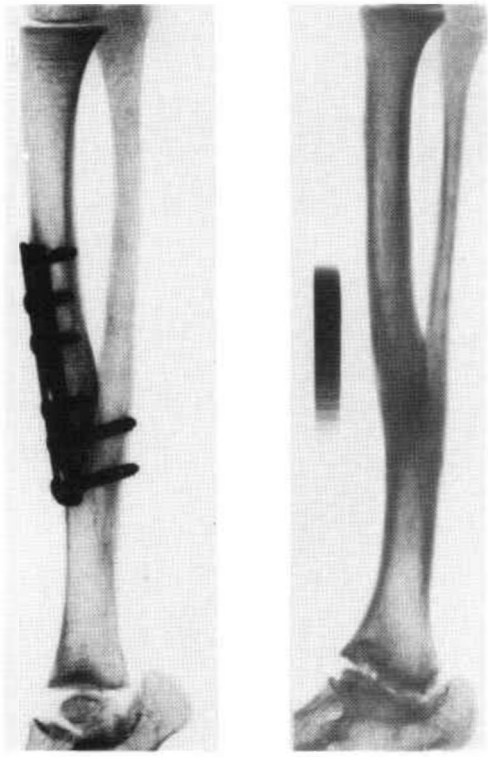
If the malformation of the foot is unilateral and it is certain from the beginning that lengthening of the leg is not recommended, the foot is brought into an equinus position, so as to allow proper fitting of a shoe or extension prosthesis.

The fusion of the tibia and the distal fibular section is performed about 3–4 months later. The distal tibial end is exposed and prepared. The fibular diaphysis is sectioned at the predetermined level. A space is then created, which is as large as possible, in the soft tissue between the two lower leg bones, for moving the distal fibular fragment to the tibial fragment. The bones are then fixed in their position by a selfcompressing plate.

Case reports

Figure 1 shows a partial tibial aplasia, with a severe deformity of the foot. Repositioning of the foot was completed at the age of 3 years, with fibula/tibia fusion being performed 1 year later. Figure 4 clearly demonstrates the recess in the calcaneus. There are no signs of damage to the large epiphyseal plate.

Meanwhile, the epiphysis is tightly fused with the residual talus and calcaneus, with the epiphyseal plate beginning to ossify. The hypertrophy of the distal tibial sections is



(a) (b)

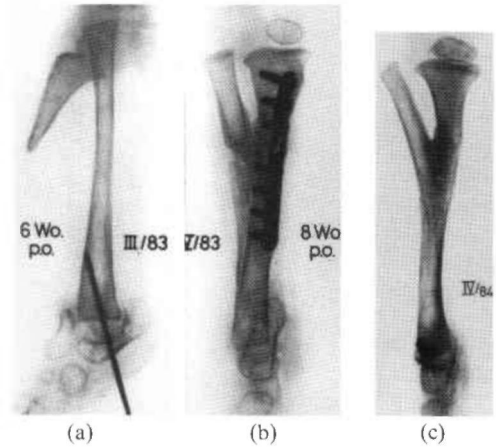
Fig. 4
a). Repositioning the foot at the age of 3, fusion completed 1 year later.

b). 10 years after repositioning the foot and 9 years after fibula/tibia fusion.

clearly demonstrated, showing a good example of functional correction of the foot (Fig. 4).

In another patient, the tibial stump was contained in a sort of skin bag (Fig. 5). The foot was repositioned at the age of 1 year, and the fibula/tibia fusion completed 8 weeks later. Due to the significant difference in the length of the legs, the foot was put in equinus. Initially, ortho-prosthetic fitting was rather difficult because of the laterally extending proximal fibula. However, with progressive growth of the leg, containment of the entire lower leg is no longer required, so the fibular head is no longer enclosed.

The authors apply this technique also in cases of diastases of the lower leg. The initial findings are demonstrated in Figure 6. This patient had on the right side a cleft foot, and on the left a lower leg diastasis. The distal tibia, contained in a skin bag, protruded from the lower leg.



(a) (b) (c)

Fig. 5.
a) After repositioning the foot
b) After fibula/tibia fusion.
c) 1 year after repositioning the foot and operative fusion.

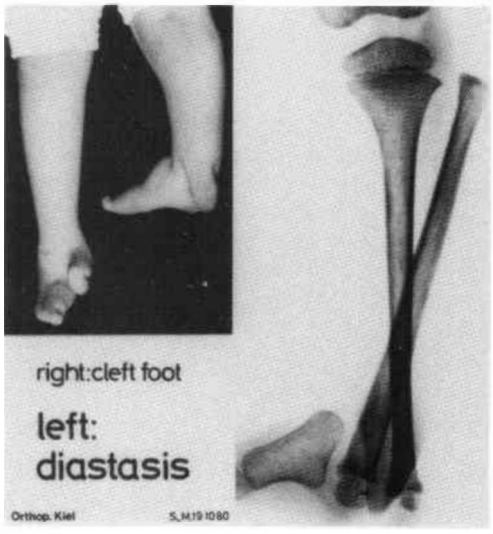
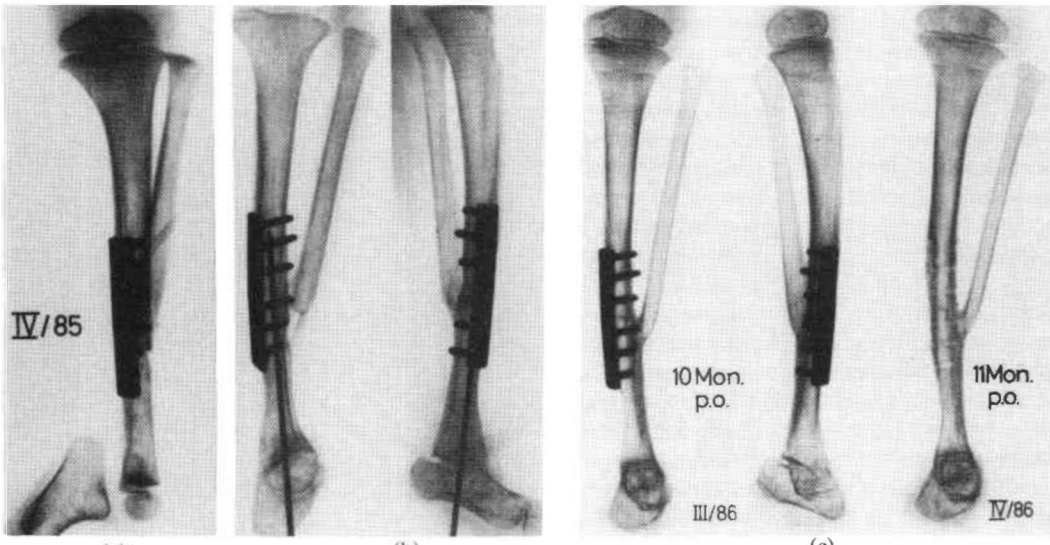


Fig. 6. Diastasis of the lower leg.

The tibia/fibula fusion was performed at a first operation and the foot was repositioned on a second occasion. Because of monodactyly, weight bearing was only possible via the repositioned calcaneus, and so the toe was amputated in a third procedure. The final state corresponds to a Pirogoff-Günter amputation. Ten weeks after the last operation the patient received the definite prosthesis (Fig. 7).

Summary

In cases of congenital partial tibial aplasia or



(a) (b) (c)

Fig 7.
a) Fibula/tibia fusion.
b) After repositioning the foot.
c) After operative fusion, repositioning of the foot and amputation of the forefoot.

so-called diastases of the lower leg, very good results are to be expected from tibia/fibula fusion in association with a repositioning of the foot. Form and function of the limb are significantly improved, with ortho-prosthetic fitting being considerably facilitated.

REFERENCES

Blauth, W., Willert, H. G. (1963). Klinik und Therapie ektromeler Mißbildungen der unteren Extremität. *Arch. Orthop. Unfall-Chir.*, **55,** 521–570.

Blauth, W. (1963). Beitrag zur operativen Behandlung schwerer Mißbildungen der Unterschenkelknochen. *Arch. Orthop. Unfall-Chir.*, **55,** 345–372.

Blauth, W. (1965). Die operative Fußunterstellung bei angeborener Tibiaaplasie. *Ärztl. Praxis,* **17,** 152.

Blauth, W. (1967). Operative Behandlung der angeborenen partiellen Tibiaaplasie. In: "Im Dienste der Chirurgie" Part 1. Glashütte (Holstein) Selbstverlag: Ethicon GmBH.

Blauth, W., Hepp., W. R. (1978). Die angeborenen Fehlbildungen der unteren Gliedmaßen. In: "Chirurgie der Gegenwart" Zenker, R., Deucher, F., Schink, W. Vol. 5. Müchen-Wien-Baltimore: Urban & Schwarzenberg.

Blauth, W., von Törne, O. (1978). Die Fibula-pro-Tibia-Fusion (Hahn-Brandes-Plastik) in der Behandlung von Knochendefekten der Tibia. *Z. Orthop.*, **116,** 20–26.

Bose, K. (1976). Congenital diastasis of the inferior tibiofibular joint: report of a case. *J. Bone Joint Surg.*, **58A,** 886–887.

Exner, G. (1967). Die Behandlung des kongenitalen Tibiadefektes durch die Hahn'sche Plastik (Translokation der Fibula). *Z. Orthop.*, **103,** 193–198.

Henkel, L., Willert, H. G. (1969). Dysmelia – a classification and a pattern of malformation in a group of congenital defects of the limbs. *J. Bone, Joint Surg.*, **51B,** 399–414.

Matthews, W. E., Mubarak, S. J., Carrol, N. C. (1977). Diastasis in tibiofibular mortise, hypoplasia of the tibia, and clubfoot in a neonate with cleft hand and cardiac anomalies: a case report. *Clin. Orthop.*, **126,** 216–219.

Milliner, S. (1978). Combined congenital diastasis of the inferior tibiofibular joint with contralateral distal femoral bifurcation and hemimelia of the medical structures. *S. Afr. Med. J.*, **54,** 531–533.

Sedgwick, W. G., Schoenecker, P. L. (1982). Congenital diastasis of the ankle joint: case report of a patient treated and followed to maturity. *J. Bone Joint Surg.*, **64A,** 450–453.

Tuli, S. M., Varma, B. P. (1972). Congenital diastasis of tibiofibular mortise. *J. Bone Joint Surg.*, **54B,** 346–350.

Wittek, A. (1906). Die operative Behandlung des partiellen Tibiadefektes. *Z. Orthop.*, **17,** 473–481.

Prosthetics and Orthotics International, 1991, 15, 131–136

Fibular deficiency and the indications for Syme's amputation

W. L. OPPENHEIM

Division of Orthopaedic Surgery, University of California, Los Angeles, USA.

Abstract

Recent literature on the subject is reviewed, and the role of Syme's amputation, reconstructive surgery and prosthetic management is discussed in relation to the severity of the condition. Amputation, which should be performed between 18 months and two years old is specifically recommended for total fibula absence with ankle instability. The operative technique is detailed.

Introduction

Congenital fibular deficiencies are frequently a source of frustration and anxiety for the physician charged with meeting both the psychosocial and functional needs of these otherwise normally functioning children. The birth of such a child can engender feelings of guilt, blame, and helplessness in the parents. The concept of ablating the foot in order to facilitate prosthetic fitting is initially not well accepted as this organ may be quite normal. The parents may question why the same technology that sends men into outer space cannot be called upon to salvage their child's limb. The physician faced with this scenario must improvise, based on his expertise in several areas including orthotics and prosthetics, reconstructive ankle surgery and leg lengthening. Considering the variability of the condition, it is little surprise that over the years many methods have evolved to address the situation. Unfortunately, for the profoundly involved limb, few methods, other than amputation and prosthetic fitting, have stood the test of time. The purpose of this discussion is to review the subject of fibular deficiency, to examine the treatments currently available, and to put the subject of amputation into proper perspective.

All correspondence to be addressed to Dr. Oppenheim, Division of Orthopaedic Surgery, University of California, Los Angeles, School of Medicine, 10833 Le Conte Avenue, Los Angeles, CA 90024–6902, USA.

Background

The fibula serves an integral purpose in the lower limb as a lateral buttress to the talus as weight is transmitted across the tibial plafond during plantigrade activity. In addition, strain determinations of the fibula, imply that it maintains a weight-bearing function during stance (Lambert, 1971). Congenital malformations of the fibula result from an alteration of musculoskeletal organogenesis that probably occurs in humans at approximately the fourth to sixth week of embryologic life. Because these malformations are not associated with classic modes of genetic transmission it is likely that they result from some embryonic insult perhaps at variable times during the development of the limb bud. Experimental evidence suggests that the earlier the insult, the more the involvement of the proximal femur and fibula. Later insults involve the fibula and foot to a greater degree (Pappas *et al.*, 1972). Whatever the timing, the range of severity is from simple hypoplasia to a total absence of the fibula often associated with proximal femoral focal deficiency, shortening and/or bowing of the tibia, general limb growth retardation, delayed epiphyseal ossification, absence of the lateral or other rays of the foot, tarsal coalitions, residual fibrous bands, deficiency of muscle, genu valgum, and loss of ankle integrity. Upper limb anomalies are seen in a significant proportion of cases (Achterman and Kalamchi, 1979; Coventry and Johnson,

1952; Herring *et al.*, 1986; Hootnick *et al.*, 1977; Westin *et al.*, 1976). Although rare, the fibula is the most commonly congenitally absent long bone (Coventry and Johnson, 1952).

Classification

From a clinical standpoint the most important issues are the stability and function of the ankle and foot and the overall length of the limb. To better characterise the variability, a number of classifications have evolved. Coventry and Johnson (1952) subdivided the condition into three types based on severity. In Type I only one limb is affected and the degree of deficiency ranges from a shortened fibula to partial absence of its upper portion. In Type II, the involvement is unilateral but absence of the fibula is complete. In Type III, the involvement may be bilateral, and the deformity may occur in association with other malformations such as proximal femoral focal deficiency. Recently Achterman and Kalamchi (1979) refined Type I into Type I-A and Type I-B. In Type I-A, the entire fibula is present, but is shortened with the proximal fibular epiphysis distal to the upper physis of the tibia and the distal fibular physis proximal to the dome of the talus. In Type I-B there is partial absence of the upper fibula, and distally the lower fibular epiphysis is elevated above the talar dome and thus not buttressing the ankle. Generally, in Type I deformities the foot remains plantigrade but may be associated with a ball and socket ankle joint; equinovalgus or equinovarus may also be present. While some surgeons have commented that the degree of ankle instability is related to the absence of rays (e.g., a four toed foot can be salvaged, but a three toed foot should be sacrificed), the author has found that ankle stability can be directly assessed and that a three toed foot can often be quite serviceable. Thus, most Type I feet will not be converted to amputations. In Type II, however, the entire fibula is absent, the incidence of tibial bowing frequent, and ankle instability is the rule. In classifying these anomalies, the degree of tibial shortening increases with advancing staging, though associated femoral shortening is maximal in the Type I category (Achterman and Kalamchi, 1979).

In Type II deformities due to the lack of lateral malleolar buttress and lateral ligamentous support of the ankle joint, the foot very often tends to become subluxed into a valgus and equinus position. This can be accentuated by a fibrous anlage of bone that extends from the upper end of the tibia to the calcaneus, exerting a tethering force on the tibia. It is in these cases that surgical salvage of the limb is most difficult. In areas where cultural concerns do not allow amputation, or in lesser developed areas where surgery may not be readily available, it is both possible and permissible to fit a prosthesis around the foot. Surgically, from the turn of the century until the 1950's attention was directed toward stabilisation of the ankle followed by leg length equalisation. Stabilisation of the ankle was first described by Braun (1886). Since then a number of different methods have come and gone, many of which are now recognised as counter-productive. For example, stabilisation by arthrodesis was complicated by delayed ossification of the epiphyses, as well as by inadvertent injury to the distal tibial physis. The Bardenhauer procedure (Rincheval, 1895) involved inserting the talus into a sagittal split in the tibia, clearly breaching the growth plate. The Albee (1921) procedure involved the substitution of autogenous bone grafted into the tibial metaphysis in an effort to replace the fibular malleolus. With growth of the tibia, the buttress so created rose above the level of the ankle. More recent attempts at ankle stabilisation have centred on the Gruca procedure (Gruca, 1959; Serafin, 1967). In a review of this technique by Thomas and Williams (1987) at the Royal Children's Hospital in Melbourne patients were considered for the procedure only if they exhibited minimal shortening of the tibia, and a foot that comfortably reached the ground with no gross deformity. The operative technique consisted of an oblique sectioning in the sagittal plane at a point between the lateral and middle thirds of the lower articular surface of the distal tibial epiphysis and extending obliquely and medially for about 7 cm. The medial tibial fragment was then displaced upward and medially 1.5 cm, and the gap between the fragments above the physis packed with cortical bone. The space at the site of the growth plate was filled with adipose tissue to prevent physeal bridging and allow further growth. The tibial fragments were transfixed by two screws to maintain positioning during consolidation. A

primitive ankle mortise was thus formed. Of seven attempts reported, three have been converted to an amputation, three were awaiting amputation, and the remaining patient had embarked on a programme of leg length equalisation. The study concluded that although in some patients a satisfactory ankle may be achieved, this procedure was not suitable for every patient, and in most cases the operation can only be regarded as an interim procedure to be followed by a Syme's amputation due to the progressive leg-length inequality that develops.

Ankle disarticulation and prosthetic fitting

In approaching the subject of conversion to a prosthetic situation, Syme's amputation should be regarded as a reconstructive rather than as an ablative procedure. Kruger and Talbott (1961) pointed out that while the goal of preserving the foot might be laudable, in fact many children underwent repetitive operations culminating eventually in Syme's amputation, and that upon honest review there was good evidence that the procedure should have been offered as a primary treatment rather than for secondary salvage. Similarly, the author (Anderson *et al.*, 1984) found a higher incidence of complications in a series of 61 patients at the Los Angeles Shriner's Hospital when Syme's amputation was performed as a salvage procedure in multiply operated limbs rather than as a primary procedure. In this series with the exception of one patient who died of unrelated causes, all patients were ambulatory, and patient satisfaction was excellent. Patients participated in sports activities including bicycling, swimming, football, soccer, and roller skating. Patients did report occasional problems such as callouses and rashes, but on closer inspection these problems seemed to be related to prosthetic fit rather than to the stump *per se* and were easily addressed with minor prosthetic accommodations in the socket. Posterior heel pad migration was commonly encountered though it rarely required surgical intervention. Hypertrophy of the skin over the distal tibia, and prosthetic adjustment were usually enough to compensate for changes in heel pad position. Forty percent of the patients felt that they had no functional restriction at all and all of the adults reviewed were employed. In another study evaluating the physical and psychological function in young patients after a Syme's amputation, the results demonstrated a surprisingly easy adjustment process (Herring *et al.*, 1986.) Patients were able to compete in most athletic endeavours, and the prosthetic device provided a cosmetically acceptable appearance. These authors noted, however, that the age of amputation was of major importance, and that it was preferable that the procedure be done between the ages of 18 months and two years since at this time the infant has an incompletely developed body image and adapts to the new physical status quite quickly. It seems that a missing foot compensated by a functional prosthesis is more acceptable to a child or teenager than a significantly deformed foot which compromises activities and gait. Further enhancing the activities of these amputees are recent advances in energy storing prosthetic feet utilising newer materials which are now available and which have further extended their abilities in terms of athletic competition; they also report a more lively feel to the leg (Burgess *et al.*, 1983; Wagner *et al.*, 1987). In brief, particularly in the severely involved child, it is more definitive and simpler to lengthen the prosthesis each year rather than lengthen the child. Young children growing up with such a conversion are fully active and functional (Anderson *et al.*, 1984; Herring *et al.*, 1986).

Indications and technique of ankle disarticulation in children

James Syme first described ankle disarticulation with preservation of the heel pad in 1843, writing that "the risk to life would be small, that a more comfortable stump would be afforded, and that the limb would be more seemly and useful for progressive motion". Since then the procedure has been modified, particularly for the child, though the basics remain.

The indications for a Syme's amputation are (1) a deformity of the foot so severe that any surgery to make the foot plantigrade and functional is likely to fail (Wood *et al.*, 1965) and (2) a leg-length discrepancy of 7.5 cm or more, actual or predicted, by the time of skeletal maturity (Thomas and Williams, 1987). This value of 7.5cm is somewhat arbitrary and tends to vary between physicians (Pappas *et al.*,

1972; Thomas and Williams, 1987). The author now makes his decision more based on the ankle situation than the leg length discrepancy, provided the discrepancy at growth is not projected to exceed a 15 cm combined femoral and tibial discrepancy.

The procedure of Syme's ankle disarticulation begins by marking out the anterior and posterior skin flaps and carrying the dissection down through the subcutaneous tissue to the level of the medial and lateral collateral ligaments of the ankle. These ligaments are identified and divided so that the talar dome can then be pulled forward away from the distal tibia. In the interval between the talus and tibia the extensor hallucis longus tendon is identified as a key to the location of the neurovascular bundle posteromedially. By protecting this tendon and drawing it medially with a retractor, the dissection can be continued with the neurovascular bundle well-protected. The calcaneus is excised from the heel pad in a *subperiosteal* fashion so that the periosteum remains thus maintaining the hydraulic structural function of the fat pad. The entire calcaneal apophysis is excised so that it will not persist as an ossicle later in growth. Two centimeters of the Achilles tendon is resected so that there will be no tendency to reattach and pull the heel pad posteriorly. Once the talus and calcaneus and the remainder of the foot have been excised, the distal end of the tibia is then shaved with the use of a knife creating a broad base for weightbearing with or without a prosthesis but taking special care not to injure the distal tibial physis. This is done only in older children since smoothing and remodelling occur spontaneously in the younger child once the talus no longer occupies the mortise. The heel pad is then stabilised underneath the tibia utilising a K-wire and the skin is closed over a Penrose drain with interrupted nylon stitches for the skin. In approximating the heel pad to the tibia care is taken to maintain the neurovascular bundle all the way to the tip of the flap and to avoid crimping. After skin repair a spica cast is applied for short stumps (as when the amputation is combined with a knee fusion for PFFD) and a long leg cast applied when the knee can be effectively bent to prevent the cast from coming off. The drain is removed at two days, and six weeks is allowed for soft tissue healing. By eight weeks the stump is generally ready for prosthetic fitting.

Some authors have reported problems with stabilisation of the heel pad and have preferred the Boyd amputation in which the calcaneus is trimmed and displaced anteriorly in an effort to fuse it into the distal tibia or at least to stabilise it under the tibial plafond (Blum and Kalamchi, 1982; Boyd, 1939). The author has some experience with this procedure, and feels it is a reasonable alternative. There is danger to the distal tibial physis in attempting to expose the epiphyseal ossification centre, especially in this group of children in which the ossification can be delayed and the nucleus, at the time of surgery, quite small.

Alternate approaches

In cases of Kalamchi Type I hypoplasia in which ankle stability is not a problem, or in which the ankle can be made stable and plantigrade either by fibula lengthening or supramalleolar osteotomy, the secondary problem of equalising leg lengths becomes paramount. For mild discrepancies no treatment, a lift, or contralateral epiphysiodesis will suffice. Several authors have documented that growth inhibition was constant throughout childhood and that the relative difference in leg lengths remains constant (Hootnick *et al.*, 1977· Moseky, 1977; Ring, 1959; Westin *et al.*, 1976). Thus a 10% discrepancy at birth will translate to 10% discrepancy at growth, but the absolute amount of shortening will of course increase. For an average 37 cm tibia, this would translate to 3.7 cm. It is important to remember that the femur is also short, and that the total amount of shortening in the limb is the sum of all of the regions of shortening. The foot itself may also be short and this should be taken into account as well in planning future treatment. In general, total discrepancies predicted to be over 5 cm at growth are usually addressed by leg lengthening as opposed to shortening techniques. Although the newer methods of lengthening — Wagner, Ilizarov, DeBastiani — are reporting extended lengthenings in areas previously thought to represent contraindications (Bjerkreim and Hellum, 1983; Dal Monte and Donzelli, 1987; DeBastiani *et al.*, 1986; Paley, 1988), a word of warning is in order. In congenital conditions, as opposed to acquired conditions, the limb is programmed to be short. Bringing such a limb out to the length of the opposite member amounts to over-lengthening not only the bone but the soft tissues as well. Under these

circumstances small associated deformities such as genu valgum, or ankle subluxation may be accentuated by the act of lengthening itself. Several lengthenings may have to be done throughout growth to "keep up" and the average time of disability and recovery may be in the order of 12–18 months per segment lengthened. This is particularly true in the tibia where leg-lengthening is frequently tardy in healing, and may require supplemental autogenous grafts and expensive prolonged and repeated hospitalisations. The economic and psychologic burden on both the family and the child undergoing the lengthening procedure must also be considered. The complication rate for lengthening approaches 100%, and a marginal foot or tenuous union may require chronic bracing and preclude an active childhood (Aldeghiri *et al.*, 1985; Bjerkreim and Hellum, 1983; Dal Monte and Donzelli, 1987; Mosca and Moseley, 1986). For Kalamchi Type I the author believes that lengthening is a reasonable and attainable goal in selected cases. In Type II deformities with grossly unstable ankles, in view of the prolonged course in preserving a less than satisfactory ankle and the speculative nature of extended lengthening in congenital situations in general, the alternative of conversion to ankle disarticulation is still the primary treatment to be considered. Furthermore, if an amputation is to be done, it should be done early so the child experiences the least psychologic disturbance (Anderson *et al.*, 1984; Herring *et al.*, 1986).

Conclusion

A Syme's amputation in a young patient is compatible with athletic and psychological function closely approaching that of a non-handicapped child of the same age. It should be considered as a primary reconstructive procedure rather than a last resort in patients with total congenital fibular absence. With lesser degrees of fibular involvement each patient must be individually assessed so that the treatment best suited to his or her personal needs can be selected. In spite of the newer methods of lengthening, ankle disarticulation and/or prosthetic fitting remains the standard treatment against which the newer methods of reconstruction and lengthening must be functionally compared. Although a number of procedures are now being proposed as alternatives to a Syme's amputation and prosthetic fitting, none have yet duplicated the degree of long-term success that the Syme's procedure has enjoyed.

REFERENCES

Achterman, C., Kalamchi, A. (1979). Congenital deficiency of the fibula. *J. Bone Joint Surg.*, **61B**, 133–137.

Albee, F. (1921). Orthopaedic and Reconstruction Surgery. Philadelphia: W. B. Saunders Co., p. 914.

Aldeghiri, R., DeBastiani, G., Renzi-Brivio, L. (1985). Allungamento diafisario dell'arto inferiore (studio di 78 casi). *Chir. Organi. Mov.*, **70**, 111–119.

Anderson, L., Westin, G. W., Oppenheim, W. L. (1984). Syme amputation in children: indications, results, and long-term follow-up. *J. Pediatr. Orthop.*, **4**, 550–554.

Bjerkreim, I., Hellum, C. (1983). Femur lengthening using the Wagner technique. *Acta Orthop. Scand.*, **54**, 263–266.

Blum, C. E., Kalamchi, A. (1982). Boyd amputations in children. *Clin. Orthop.*, **165**, 138–143.

Boyd, H. B. (1939). Amputation of the foot with calcaneotibial arthrodesis. *J. Bone Joint Surg.*, **21**, 997–1000.

Braun, H. (1886). Ueber die intrauterinen frakturen der tibia. *Arch. Klin. Chir.*, **34**, 668.

Burgess, E. M., Hittenberger, D. A., Forsgren, S. M., Lindh, D. (1983). The Seattle prosthetic foot — a design for active sports: preliminary studies. *Orthot. Prosthet.*, **37**, 25–31.

Coventry, M. B., Johnson, E. W. (1952). Congenital absence of the fibula. *J. Bone Joint. Surg.*, **34A**, 941–955.

Dal Monte, A., Donzelli, O. (1987). Tibial lengthening according to Ilizarov in congenital hypoplasia of the leg. *J. Pediatr. Orthop.*, **7**, 135–138.

DeBastiani, G., Aldeghiri, R., Renzi-Brivio, L., Trivella, G. (1986). Chondrodiastasis controlled symmetrical distraction of the epiphyseal plate; limb lengthening in children. *J. Bone Joint Surg.*, **68B**, 550–556.

Gruca, A. (1959). Chirurgia Ortopedyczna, Warsaw: PZWL.

Herring, J. A., Barnhill, B., Gaffney, C. (1986). Syme amputation: an examination of the physical and psychological function in young patients. *J. Bone Joint Surg.*, **68A**, 573–578.

HOOTNICK, D., BOYD, N. A., FIXSEN, J. A., LLOYD-ROBERTS, G. C. (1977). The natural history and management of congenital short tibia with dysplasia or absence of the fibula: a preliminary case report. *J. Bone Joint Surg.*, **59B,** 267–271.

KRUGER, L. M., TALBOTT, R. D. (1961). Amputation and prosthesis as definitive treatment in congenital absence of the fibula. *J. Bone Joint Surg.*, **43A,** 625–642.

LAMBERT, K. L. (1971). The weight-bearing function of the fibula. *J. Bone Joint Surg.* **53A,** 507–513.

MOSCA, V., MOSELEY, C. F. (1986). Complications of Wagner leg-lengthening and their avoidance. *Orthop. Trans.*, **10,** 462.

MOSELEY, C. F. (1977). A straight-line graph for leg-length discrepancies. *J. Bone Joint Surg.*, **59A,** 174.

PALEY, D. (1988). Current techniques of limb lengthening. *J. Pediatr. Orthop.*, **8,** 73–92.

PAPPAS, A. M., HANAWALT, B. J., ANDERSON, M. (1972). Congenital defects of the fibula. *Orthop. Clin. North Am.*, **3,** 187–199.

RINCHEVAL (1894). Ein neues operations verfahren zur behandlung cogenitaler deffecte eines unterarm und unterschenkelknochens. *Arch. Klin. Chir.*, **48,** 802.

RING, P. A. (1959). Congenital short femur. *J. Bone Joint Surg.*, **41B,** 73–79.

SERAFIN, J. (1967). A new operation for congenital absence of the fibula: preliminary report: *J. Bone Joint Surg.*, **49B,** 59–65.

SYME, J. (1843). Amputation at the ankle joint. *London Edinburgh Monthly Journal.* **2,** 93–96.

THOMAS, I. H., WILLIAMS, P. F. (1987). Operation for congenital absence of the fibula. *J. Bone Joint Surg.*, **69B,** 587–592.

WAGNER, J., SIENKO, S., SUPAN, T., BARTH, D. (1987). Motion analysis of SACH vs. flex foot in moderately active below-knee amputees. *Clin. Prosthet. Orthot.*, **2, (1),** 55–62.

WESTIN, G. W., SAKAI, D. N., WOOD, W. L. (1976). Congenital longitudinal deficiency of the fibula. *J. Bone Joint Surg.*, **58A,** 492–496.

WOOD, W. L., ZLOTSKY, N., AND WESTIN, G. W. (1965). Congenital absence of the fibula: treatment by Syme amputation — indications and technique. *J. Bone Joint Surg.*, **47A,** 1159–1169.

Prosthetics and Orthotics International, 1991, 15, 137–139

Reconstructive surgery for fibular deficiency

O. E. SHATILOV, A. V. ROZKOV and T. V. CHEMINOVA

The Leningrad Scientific Research Institute of Prosthetics, USSR

Abstract

Three types of fibular deficiency are described which determine the nature of the surgery and prosthesis required. The surgical management of 50 patients who had a total of 103 operations is described.

Introduction

The seriousness of the pathology, variety of clinical manifestations, and the involvement of several limbs makes the medical rehabilitation of children with longitudinal congenital deficiencies very difficult. It is necessary to apply the whole complex of modern methods of surgical treatment and prosthetics and orthotics, in order to solve the problem.

Treatment must eliminate the deformity and shortening, improve the weightbearing and motor function of the limb and allow the fitment of an improved prosthesis. During the last 30 years the authors have followed up 213 children, 50 of whom underwent surgical reconstruction.

Fibular deficiency

Absence or deformity of the fibula, the most common longitudinal lower limb deficiency, was first described by Göller (a German scientist) in 1967 (Coventry and Johnson, 1962). Clinically this deficiency presents with shortening, malformation and deformity of the foot. In the majority of cases there is angulation of the distal tibia with convexity forwards and medial rotation. The foot is in equinovalgus and the ankle is subluxed laterally. The fourth and fifth metatarsals and toes are often absent. The talus and calcaneus are often deformed and may be fused. The tibial deformity and foot displacement (sometimes as far as the middle third of the leg) are caused by the presence of a fibro-cartilaginous cord or anläge representing the fibula. Haudek in 1896 was the first to describe this cord (Thompson *et al.*, 1957), and Karchinov (1963), Karimova (1975) and Bedova (1981) studied its location, structure and influence on the deformity. In addition some children have a valgus deformity of the knee with a flexion contracture.

Treatment

Three types of fibular deficiency are

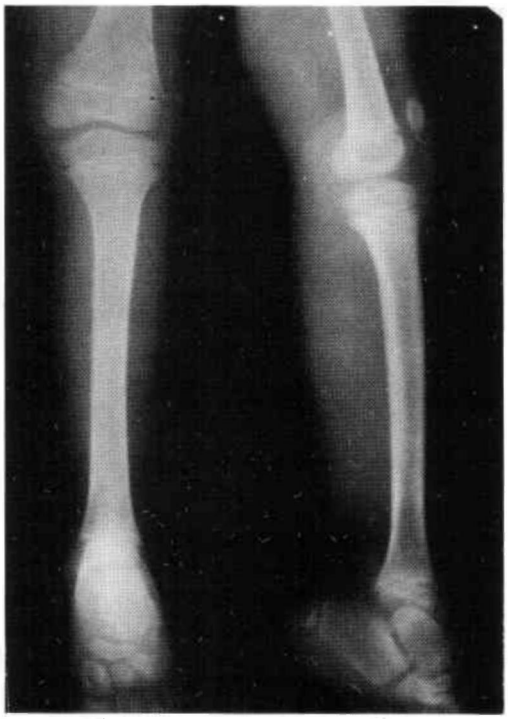


Fig. 1. An example of a Group 1 deficiency.

All correspondence to be addressed to Dr. O. E. Shatilov, The Leningrad Scientific Research Institute of Prosthetics, Bestuzlevskaya 50, Leningrad 195067, USSR.

recognised determining the nature of the surgery and prosthesis.

The first (Fig. 1) includes 10 patients with fibula absence, significant shortening, outward subluxation of the equinus foot, but with no tibial deformity. In these patients tenoligamentocapsulotomy was performed, and the corrected position maintained in plaster. No fibrus anläge was found at operation. A leg lengthening procedure using hinge-distracting apparatus was subsequently performed on 5 children.

The second group (Fig. 2) of 35 children had absence of the fibula accompanied by tibial angulation and rotation, and an equinovarus deformity of the foot which was subluxed backwards and outwards. All had a fibrous anläge and a flexion deformity of the knee was often seen. A hinge-distracting apparatus was used in the first operation to overcome the knee joint contracture and thus lengthen the limb. In a second stage the fibrous cord was excised, tendons elongated and the foot repositioned after tenotomy and exposure of the joint capsule. The resulting skin defect on the back of the leg was closed by a tube pedicle graft (Fig. 3).

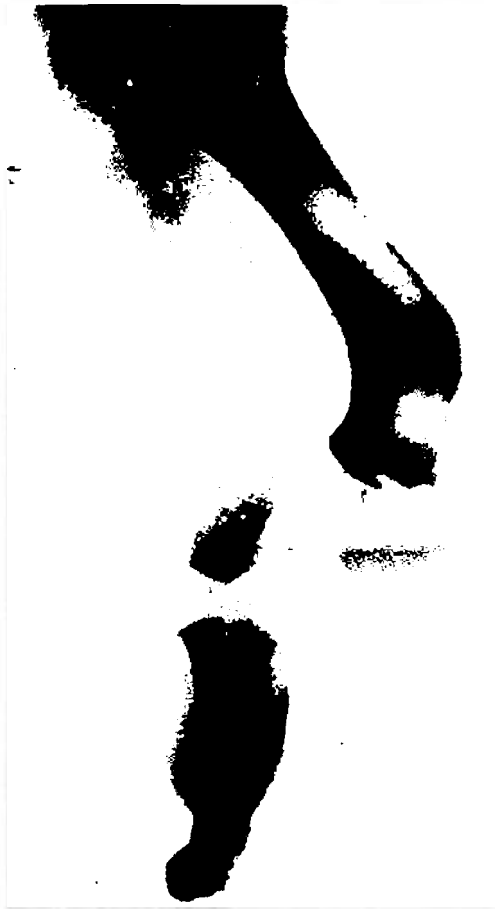


Fig. 2. Group 2 deficiency.

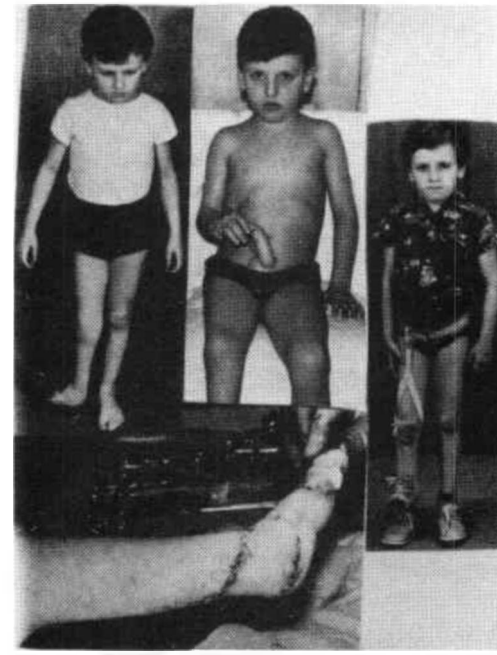


Fig. 3. Preoperative appearance, closure of skin defect using pedicle graft and extension prosthesis.

In 18 children the foot was repositioned using a distraction apparatus. A supracondylar

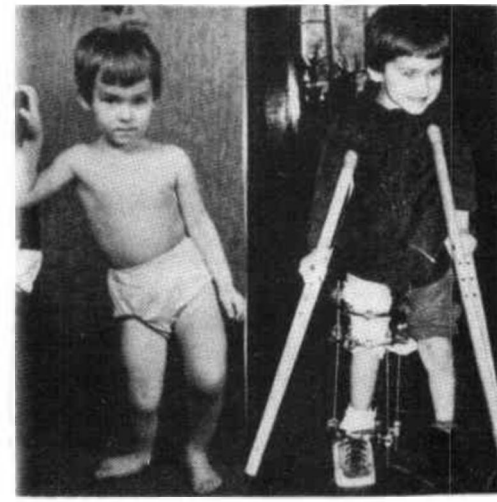


Fig. 4. Child undergoing lengthening procedure.

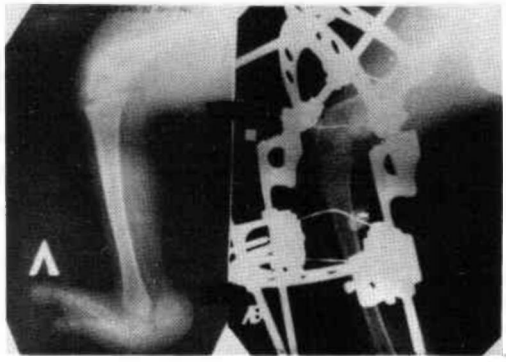


Fig. 5. Group 3 deficiency showing knee abnormality and hinge distraction apparatus.

osteotomy of the femur or correction of the tibia was necessary in 12 patients in order to improve the alignment of the limb. Femoral lengthening was carried out in 7 cases, tibial lengthening in 12 and 3 cases needed lengthening of both bones (Fig. 4). Femoral lengthening of 6–10 cm was achieved at the expense of the distal epiphysis. In most cases tibial lengthening of 4–16 cm was at the expense of the proximal epiphysis. In all cases the Ilizarov apparatus was used for distraction. Two children required repeated lengthening and three subsequently had a Pirogov type of amputation during adolescence.

The third group (Fig. 5) contained those whose absence of the fibula was accompanied by ankylosis of the knee in flexion. The presence of the epiphysis made it possible to correct the deformity and to increase the length with the aid of a distraction apparatus.

Study of the long term results showed that the corrected foot position was preserved. Tibial torsion did not increase, indeed it actually decreased in some young children. In 4 cases the flexion contracture of the knee recurred. The lengthening was preserved and limb growth continued in all children.

The variety of the manifestations, their combination with joint deformities, and their effect on the overall development of the child requires that surgery and provision of prostheses start early. In carrying out reconstructive procedures, it is important to follow the steps in sequence and to take into account the possibility of providing a suitable prosthesis between the stages and after the surgery.

It is desirable to start by correcting contractures of the proximal joints, and excising the fibrous cord and correcting the subluxation of the foot, and thus increasing the length of the limb. The use of the hinge-distraction apparatus makes such corrections easier.

REFERENCES

COVENTRY, M. B., JOHNSON, E. W. (1962). Congenital absence of the fibula. *J. Bone Joint Surg.*, **34A**, 941–956.

DEDOVA V. D., LAURISHCHEVA, G. I., AMETOVA, I. K. (1981). O fibrozom tiazhe pri vrozhdennom otsutstvii malobertsovoi kosti. *Ortop. Travmatol. Protez.*, **9**, 22–27.

KARCHINOV, K. (1963). Vestnik Khirurgii, **8**, 98–103.

KARIMOVA, L. F., KNUGE, V. (1975). Rannya diagnostika i lechenia ortopedicheskikh zabolevanii ou decteii. 66–68.

THOMPSON, T. C., STRAUB, L. R., ARNOLD, W. D. (1957). Congenital absence of the fibula. *J. Bone Joint Surg.*, **39A**, 1229–1237.

Prosthetics and Orthotics International, 1991, 15, 140–142

The care of the limb deficient child in Australia

L. E. JONES* and A. LIPSON†

**Royal South Sydney Hospital, Zetland, Australia*
†Royal Alexandra Hospital for Children, Camperdown, Australia

Abstract
The incidence of limb deficiency is given as is the prevalence of prosthetic use in Australia. The organisation of clinics and the Free Limb Scheme is described and its effects discussed.

Epidemiology

There are no precise figures on the number of children in Australia with congenital or acquired limb deficiencies. The incidence of perinatal limb reduction defects has been shown to be 50 per 100,000 births and is broadly similar in each State (University of Sydney, 1986). It should be noted that this figure includes perinatal deaths. Many are finger deficiencies and syndactyly and are not reviewed in a limb deficiency clinic. The prevalence of prosthetic use in Australian limb deficient children is 14.88/100,000 (Jones, 1988). At the Royal Alexandra Hospital for Children in Sydney 25-30 new patients with congenital limb deficiency are seen each year. Approximately 30 per cent of these children will require prosthetic treatment. The ratio of congenital to acquired limb deficiency among prosthetic users is 3.7:1. The vast majority are still congenital, particularly the wearers of Syme's and PTB prostheses are congenital limb deficient children who have undergone surgical conversion. The causes of limb deficiency in New South Wales children who are prosthetic users are shown in Figure 1. There is a preponderance of males with a sex ratio of 1.8 males to 1 female.

All correspondence to be addressed to Dr. Lorraine Jones, Staff Specialist in Rehabilitation Medicine, Royal South Sydney Hospital, Joynton Avenue, Zetland, NSW 2017, Australia.

Limb deficiency clinics

Limb deficiency clinics are run in the major children's hospitals in the capital cities of each State. These clinics are run by a paediatrician/dysmorphologist in Sydney, an orthopaedic surgeon in Melbourne and a rehabilitation physician in Brisbane. In the other cities, children are seen in the major amputee clinics, though at a separate time from the adult patients. The Royal Alexandra Hospital for Children Clinic in Sydney is a multi-disciplinary clinic with attending staff members, including a paediatrician/dysmorphologist, an orthopaedic surgeon, a rehabilitation physician, an occupational therapist, a physiotherapist, a prosthetist and a social worker.

The Brisbane clinic contains a rehabilitation physician, a physiotherapist, occupational therapist, but no paediatrician or orthopaedic surgeon. They are available for consultation. Infants and children are referred to the clinics by paediatricians and general practitioners and orthopaedic surgeons.

Standard international guidelines are used for early prosthetic fitting of both upper and

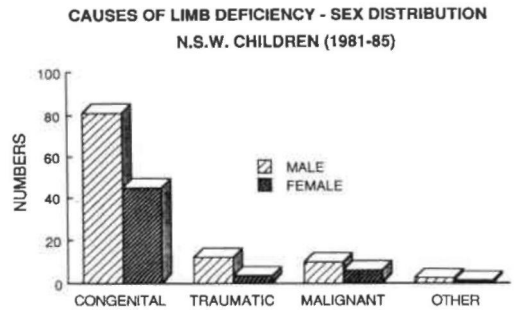


Fig. 1. Causes of limb deficiency in New South Wales.

lower limb prostheses. Upper limb prostheses are provided when the child commences bimanual activities. Lower limb prostheses are provided when the child commences to stand. Attendance at the clinic is determined by the needs of the child and the family.

Surgery and the limb deficient child

In Sydney and in Melbourne, the paediatric surgeon is an integral part of the limb deficiency clinic. His/her attendance at each clinic means that planning of surgical conversion occurs at the appropriate time after full consultation with parents and child. Tibial deficiency is treated by knee disarticulation at 3–5 years of age. Syme's amputation for fibular deficiency is performed at 3–5 years of age. A child with proximal focal femoral deficiency is not treated with rotationplasty, (the Van Nes procedure) but is fitted with an appropriate prosthesis incorporating the foot with a knee joint for the short, older child who may have a Syme's amputation for associated fibular deficiency.

Social integration of limb deficient children

Limb deficient children are integrated into the school and sporting life of ordinary childhood. Hospitalisation, when it occurs, is kept as brief as possible. Children attend normal schools and are encouraged to play in normal social and sporting clubs. There is also an active amputee sporting association for children. It was noted in an analysis of below-elbow prosthetic usage in the Free Limb Scheme that adolescents in Australia, as in other countries, frequently discard their prostheses even if they have been good prosthetic users.

Financial considerations

Attendance at limb deficiency clinics and medical/paramedical treatments are covered by universal health insurance. These clinics are subsidised by each State Government. There is in Australia a "Free Limb Scheme" which provides free prostheses to Australian residents who require them, within certain constraints. Components such as energy storing feet and hydraulic knees are not covered by the Free Limb Scheme and must be paid for by the patient. Myoelectric hands are not provided by the Scheme. The Variety Club of Australia, a charity, provides a limb bank of myoelectric hands at the Sydney clinic and shares the prosthetic costs with the Free Limb Scheme for Australian residents. Myoelectric prostheses are provided for children over the age of four, who are good prosthetic users, with a committed family, normal development, and within a reasonable geographic distance from the clinic. Body powered prostheses are not supplied if a myoelectric unit is fitted. Prostheses, in general, are renewed as often as is required depending on wear and tear and the growth of the child.

Paediatric prosthetic use

An analysis of the prosthetic usage of children under the Free Limb Scheme (1981-1985) showed that 69% of the child prosthetic users used lower limb prostheses (Jones, 1989). The prosthetic usage of these children is shown in Table 1. Upper limb users form 31% of the paediatric prosthetic population, and of these 73% are below-elbow prosthetic users.

Geographic constraints on prosthetic use

Whilst prosthetic cost is not a problem because of the Free Limb Scheme, geographic isolation of some children in decentralised States, such as Queensland, means that fewer upper limb prostheses are prescribed than those for the lower limb. In very hot areas, sweating and overheating are a problem in prosthetic usage. Children in remote areas, thousands of kilometres from a prosthetic clinic, will learn to manage without an upper limb prosthesis,

Table 1. Lower limb prosthetic use — Australian children aged 0–14 (1981-1985)*

Lower Limb	Children	Prostheses	Prostheses/ Child
Hip disarticulation	5	11	2.2
Above-knee:			
(suction)	28	65	2.3
(non-suction)	54	152	2.8
Knee disarticulation	9	18	2
Below-knee:			
(PTB)	103	348	3.4
(thigh lacing)	13	39	3
Ankle disarticulation			
(Syme's)	68	205	3
Level not classified	96	341	3.6
TOTAL	376	1,179	

* Table 1 is published with the consent of the *Australian Paediatric Journal.*

whereas the value of a lower limb prosthesis is so obvious to parents that they will ensure that the child attends clinics and that the necessary prosthesis is provided.

Summary

The care of limb deficient children is well organised in Australia at minimal cost to the parents. Prostheses are supplied when required and surgical intervention is arranged when appropriate.

REFERENCES

JONES, L. E. (1988). The Free Limb Scheme and the limb deficient child in Australia. *Aust. Paediatr. J.*, **24**, 290–294.

JONES, L. E. (1989). Prosthetic limb use in Australia 1981–1985 under the Free Limb Scheme. *Prosthet. Orthot. Int.*, **13**. 76–81.

UNIVERSITY OF SYDNEY (1986). Congenital Malformations Australia, 1981–1984 — Sydney: University of Sydney, National Perinatal Statistical Unit.

Prosthetics and Orthotics International, 1991, 15, 143–145

The care of the limb deficient child in India

V. P. SHARMA

King George Medical College, Lucknow, India

Abstract

The problem of treating limb deficient children in India is compounded by many factors, social, cultural and economic. Few attend early and most therefore manage without prosthetic care.

Introduction

India is a vast country. More than 80% of its population live in inaccessible rural areas. There is marked diversity in culture, religion, education and awareness amongst the population. But since 1981, the "Year of the Disabled", there has been a growing concern about the disabled population in the country. Till today a lack of awareness amongst the treating surgeons and the limb deficient patients has further complicated the problem. Even those children who have been provided with orthoses and prostheses are not using them regularly. Is this because the population is ignorant and do not realise what is good for them, as is being affirmed by many specialists, or is there a possibility that the appliances are not suitably designed for them? If the appliance actually helps, it would certainly be used. After all it is the user who knows best what is good for him or her.

Rehabilitation of limb deficient children

In India mostly limb deficient children do not reach the rehabilitation institutions early with the result that they adjust themselves in such a way that they can perform most activities. Social, cultural and economic constraints restrict the use of surgery to the correction of deformities. By the time they reach an institution they are grown up and unwilling to undergo surgical corrections.

Designs of aids for limb deficient children is a very complex business. It not only calls for a more scientific approach but a much better understanding of Indian Society, its culture, its gross economic disparities and its stratified structure. Any solution needs to be cost effective.

At the Rehabilitation and Artificial Limb Centre, Department of Physical Medicine and Rehabilitation, Lucknow 11,590 new cases attended the outpatient department in 1989. Out of these only 36 cases had limb deficiencies present at birth while more than twice that were of other musculoskeletal anomalies present at birth. Lower limb deficient patients were either walking without any aid or with the help of a bamboo stick. In most of the patients associated multiple congenital anomalies were present. Most of those with upper limb deficiencies were working with the normal limb and using the deficient limb just for support. Most of the patients were uneducated and engaged in menial work. Few of them were graduates seeking a job. Even the well educated patients were not willing to undergo reconstructive surgery to improve their function. All the limb deficient patients were from the poorer class.

Aetiological factors

In India various aetiological factors have been invoked; the use of contraceptive drugs before the birth of the affected child, jaundice or malaria during pregnancy. The most important factor had been exposure to a solar eclipse during pregnancy. Exact pathogenesis is not known but in this country because of religious belief, pregnant mothers are advised to avoid exposure during a solar eclipse. In one

All correspondence to be addressed to Dr V. P. Sharma, Department of Physical Medicine and Rehabilitation, King George Medical College, Lucknow 226018, India.

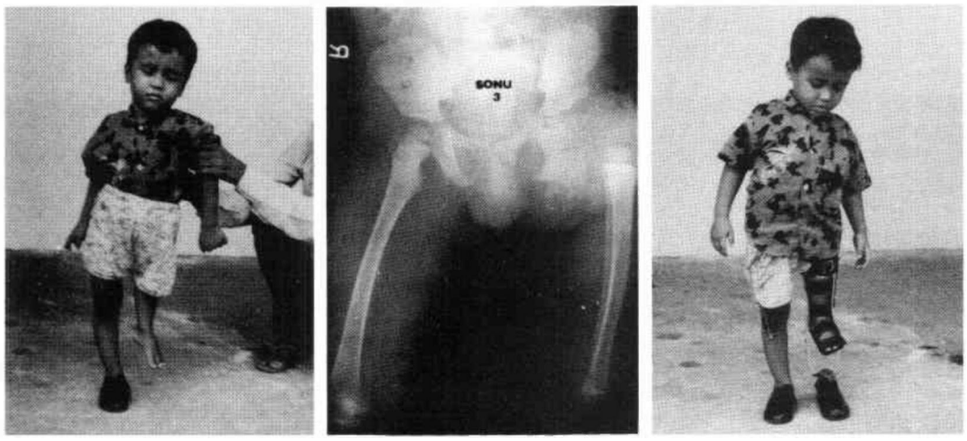


Fig. 1. A 3 year old child with longitudinal femur total deficiency and 5th ray total deficiency. Child was walking with left lower limb keeping right knee flexed. History of solar eclipse exposure during pregnancy present. Extension orthosis was fitted and now the child is walking comfortably

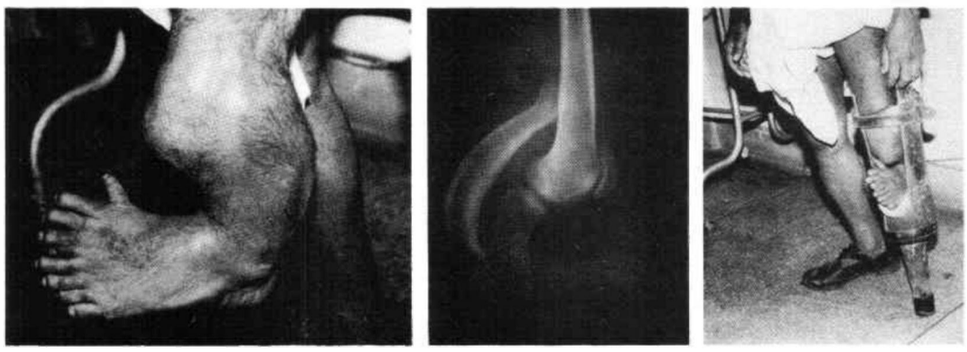


Fig. 2. A 45 year old male. Educated up to 5th standard. Farmer by occupation. Using indigenous prosthesis for last 10 years, initially he was using crutches. He can walk up to 1km a day with no difficulty.

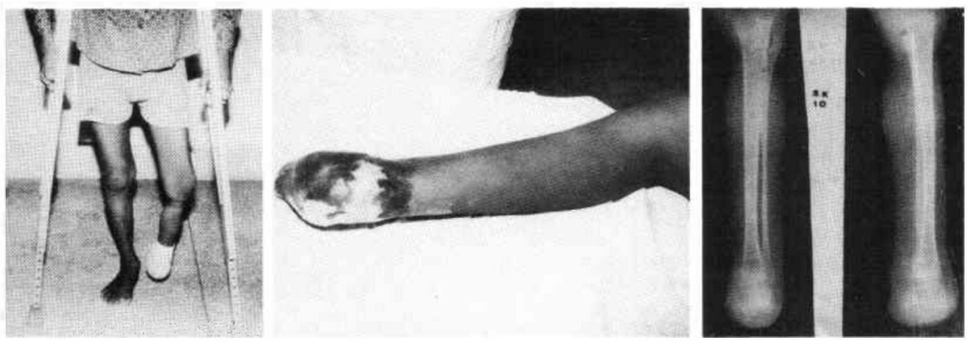


Fig. 3. A 10 year old male child uneducated having transverse tarsus partial deficiency. He has unstable scar over stump and uses crutches for walking.

study out of 30 cases 3 had definitive exposure to the solar eclipse. In the rest of the cases no clear cut history of eclipse exposure was available.

Summary

Thus in India the problem of congenital limb deficiency is entirely different to that in the western world. Most patients manage very well without any aids or appliances. However, with the fitting of a prosthesis or orthosis there had been a definitive improvement. As patients are poor, prostheses and orthoses are being provided free of cost through the help of voluntary agencies and the Ministry of Welfare, Government of India scheme. Surgical procedures are being carried out free of cost in Government Hospitals.

REFERENCES

AGARAWAL, A. K., SHARMA, V. P. *et al.* (1984). A clinical study of congenital limb deficiencies. *J. Rehabil. Asia,* **25,** 33–43.

Prosthetics and Orthotics International, 1991, 15, 146–151

The care of the limb deficient child in Japan

S. KAKURAI* and M. KIDA†

* *Teikyo University Ichihara Hospital, Ichihara City, Japan*
† *Teikyo University School of Medicine, Ichihara City, Japan*

Abstract
The incidence as well as the current state of the management of the limb deficient child in Japan is described.

General medical and welfare services in Japan

The medical service system in Japan is well organized and includes the diagnosis, treatment and follow-up care of limb deficient children. Once the child has been diagnosed as physically disabled due to limb deficiencies and/or congenital malformations, he or she has the right to receive welfare assistance under the Law for the Welfare of Physically Disabled Children. They are registered and issued with a handbook for identification and access to services. The welfare offices are in charge of the manufacture and distribution, as well as the repair, of the prosthetic appliances. Unfortunately, electric arm prostheses are not included in the inventory at the present time. Those children for whom surgical conversion seems appropriate are evaluated for short-term hospitalization at the medical agencies designated by the Ministry of Health and Welfare. However, not every facility is experienced in the management of such children, as will be shown later in the results of a questionnaire survey.

Incidence of limb deficiencies present at birth and activities of parents organizations in Japan

Thalidomide embryopathies were prevalent during the early 1960's in Japan and 306 patients (171 males and 135 females) were registered (Kida, 1987).

All correspondence to be addressed to S. Kakurai, Department Physical Medicine and Rehabilitation, Teikyo University Ichihara Hospital, 3426–3, Anegasaki, Ichihara City 299–01, Japan.

A survey has been conducted by the Japan Physician's Association for Maternal Welfare (JAMW) for monitoring birth defect babies since 1982. Approximately 120,000 new-born babies have been examined annually at 250 gynaecological hospitals and facilities throughout the country and the results registered with the International Clearing House for Birth Defects. Monitoring System (ICBDMS). According to the statistical study of Kida (1989), new-born children with limb-reduction deformity are born at a rate of 7 per 10,000 births; therefore, approximately 1,000 affected infants are born annually. Table 1 shows the frequency of limb-reduction deformities recorded by the JAMW between 1978 and 1986.

The Association of Parents of Children with Congenital Malformation of the Limbs (APCCML) was organized in 1979 and more than 1,000 families are joint members. Since the establishment of the association one of the authors has been engaged in 66 screening surveys throughout the country and has examined a total of 1,441 cases (Kida, 1989). Table 2 shows a comparison of results of limb deficiencies present at birth registered by the JAMW and screening surveys done at the APCCML. The APCCML have many activities for the psychological support of its members and also to encourage the development of social understanding.

Apart from this study, the authors wanted to investigate the situation of congenital limb deficient children who had been hospitalized in facilities for crippled children and at children's hospitals throughout the country. Some 79 questionnaires were sent and 49 answers received (62% response ratio); 23 facilities had never experienced such cases during the past

Table 1. Frequency of limb reduction deformities registered by the JAMW (1978-86) ( ) indicates per 10,000 births

	1978–81	1982–86	Total	%
Oligodactyly/finger	94 (1.70)	128 (1.92)	222 (1.82)	28.1
Oligodactyly/toe	53 (0.96)	50 (0.75)	103 (0.84)	13.0
Hand defect	14 (0.25)	—	14 (0.25)	1.8
Split hand	—	19 (0.29)	19 (0.29)	2.4
Split foot	—	9 (0.14)	9 (0.14)	1.1
Phocomelia	10 (0.18)	10 (0.15)	20 (0.16)	2.5
Transverse amelia/upper limb	—	20 (0.30)	20 (0.30)	2.5
Transverse amelia/lower limb	—	18 (0.27)	18 (0.27)	2.3
Hypoplasia/upper limb	94 (1.70)	85 (1.28)	179 (1.47)	22.6
Hypoplasia/lower limb	98 (1.77)	82 (1.23)	180 (1.48)	22.8
Constriction band syndrome	—	7 (0.10)	7 (0.10)	0.5
Subtotal	363	428	791	100.0
Total births (JAMW)	553,167	665,861	1,219,028	
Incidence of limb Reduction deformity (per 10,000 births)	6.562	6.427	6.488	
Total births (whole country)	6,457,567	7,328,382	13,785,949	
Estimated number of limb reduction deformities	4,237	4,710	8,944 [993·8/yr]	

Table 2. Comparison of results of limb reduction deformities registered by the JAMW and screening surveys done at the APCCML (Kida, 1989)

JAMW	%	APCCML Surveys	%
Oligodactyly/finger Oligodactyly/toe	28.1 13.0	Oligodactyly/finger and toe	3.9
Hypoplasia/upper limb Hypoplasia/lower limb	22.6 22.8	Hypoplasia/upper and lower limb Ulnar aplasia	30.4 1.9
Hand defect	1.8	Poland Synbrachydactyly Syndactyly/finger and toe Brachydactyly Polydactyly	8.2 4.8 1.9 1.5
Split hand Split foot	2.4 1.1	Split hand and foot	9.7
Phocomelia	2.5	Phocomelia	0.5
Transverse amelia/upper limb Transverse amelia/lower limb	2.5 2.3	Transverse amelia/upper and lower limb	1.9
Constriction band syndrome	0.5	Constriction band syndrome	26.6
		Others: Congenital multiple arthrogryposis Microtia [Total 253 Patients]	1.9 6.8

Table 3. Number of cases experienced in 26 facilities for crippled children and children's hospitals between 1988–1990

Type of Case	Number of Cases	Percentage
Unilateral upper limb	54	32.1
Bilateral upper limb	19	11.3
Unilateral lower limb	61	36.3
Bilateral lower limb	21	12.5
Unilateral upper and lower limb	7	4.2
Triple limb	3	1.8
Quadruple limb	3	1.8
Total	168	100

Table 4. Number of cases presented at childrens hospitals between 1988–1990 showing the number of surgical procedures performed and the number of prostheses supplied

Category	Unilateral cases	Bilateral cases	Total	Number of cases with surgical procedures	Number of cases fitted with prostheses
Upper limbs					
Transverse deficiencies:					
Shoulder	13	8	21	0	11 (52.4%)
Upper and forearm	17	2	19	0	12 (63.2%)
Longitudinal deficiencies:					
Complete	17	9	26	5 (19.2%)	7 (26.9%)
Partial	29	6	35	1 (2.9%)	2 (5.7%)
Total	76	25	101	6 (5.9%)	32 (31.7%)
Lower limbs					
Transverse deficiencies:					
Hip	0	2	2	0	1 (50.0%)
AK and BK	10	3	13	10 (76.9%)	12 (92.3%)
Longitudinal deficiencies:					
PFFD	7	3	10	4 (40%)	6 (60%)
Tibial:					
Complete	7	7	14	9 (64.3%)	14 (100%)
Partial	13	1	14	14 (100%)	6 (42.8%)
Fibular:					
Complete	16	3	19	6 (31.6%)	11 (57.9%)
Partial	2	0	2	0	1 (50%)
Phocomelia*	10	3	13	1 (7.1%)	12 (85.7%)
Total	65	22	87	44 (50.6%)	63 (72.4%)

* Precise classification unknown.

three years, the remaining 26 facilities had experienced a total of 168 cases (Table 3). Table 4 shows the number of cases seen, the number of surgical procedures performed and the number of cases who have been fitted with prostheses.

Fundamental treatment method for the upper limb deficient child

Among unilateral upper limb amputees in Japan — a country whose people are considered to be generally uniform in appearance and where many value cosmesis over function — most including the parents of limb deficient children, want a cosmetic hand. The split hook is seldom prescribed and used for unilateral cases because of cosmesis (Kakurai *et al.*, 1989). Conversely, bilateral cases are fitted and trained with functional hooks. Due to the small experience with externally powered arm prostheses for higher levels of amputation in this country, most amelic cases have never been fitted with functional arms or externally powered arm prostheses. However, they have been trained to be independent in activities of daily living using their feet.

Fundamental treatment method for the lower limb deficient child

The fundamental programmes for lower limb deficient children in Japan are similar to those of other advanced countries. Teenage amputees are fitted with endoskeletal lower limb prostheses; the diameter of the tubes for such prostheses is 25 mm, whereas that for adults is 30 mm. Although many adult amputees have benefited from turntables and torque absorbers in order to adapt to Japanese domestic life styles, those for youngsters are unfortunately not available at the present time.

Surgical conversion of congenital lower limb deficiencies in Japan

In order to determine the opinions of orthopaedic surgeons engaged in the treatment of crippled children, a questionnaire survey of surgical conversions was sent to them. The following are the results among these doctors of optional treatment methods for typical unilateral congenital lower limb deficiencies; the numbers in the parentheses indicate the number of responses.

Unilateral PFFD (Aitken-Type C):

Knee arthrodesis and Syme's amputation (4); Van Nes rotation osteotomy of the leg (3); hip surgery and leg lengthening (2); leg lengthening (2); hip surgery, knee arthrodesis and Syme's amputation (1); conservative treatment (3).

As the incidence of PFFD in Japan is rather low, the opinions expressed are not statistically significant.

Unilateral total tibial deficiency

Knee disarticulation (7); Brown procedure (5); Brown procedure or knee disarticulation (2); Syme's amputation or knee disarticulation (1); leg lengthening (1); below-knee amputation (1); conservative treatment (5).

It seems from the authors' experience that absolute indications for the Brown procedure are not very extensive.

Unilateral total fibular deficiency

Syme's amputation (9); leg lengthening (2); Syme's amputation or leg lengthening (1); conservative treatment (14).

Although the answers are few in number, these opinions reflect the Japanese doctors' philosophy of surgical conversion of typical lower limb-deficiencies.

Education curriculum for medical and allied health professions on the management of the limb deficient child

That part of the curriculum dealing with the management of limb-deficiency is generally unsatisfactory, with the exception of a one week course on prosthetics and orthotics held under the auspices of the Ministry of Health and Welfare. Since 1975 more than 2,000 doctors have attended this course. At the present time, only one hour of lectures by the authors is included.

Special prostheses and devices for the limb deficient child developed in Japan

Original prostheses for limb deficient children developed in Japan are few, except for the passive hand for small children (Fig. 1), special socket design for unilateral hemipelvectomy developed by R. Takahashi in which the body weight is supported by the sound ischium (Fig. 2) and a torque absorber made of titanium alloy (Betto *et al.*, 1989) (Fig. 3).

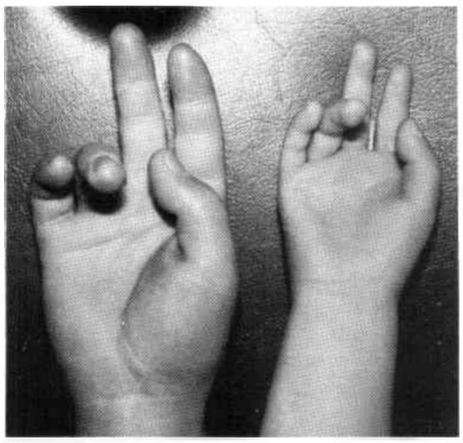


Fig. 1. Passive hand for unilateral upper limb amputated adults (left) and small children (right)

Nowadays most limb deficient children, including multiple cases, attend normal public schools. The curriculum in music of normal lower primary schools, from the third grade until the completion of compulsory education, necessitates that every child must play the recorder-flute. This is very difficult and sometimes impossible for the upper limb deficient and/or handicapped child. This has led us to the development of two kinds of recorder-flute for the unilateral arm amputee or handicapped child (Kakurai *et al.*, 1983). The first type for unilateral upper limb amputees uses a special mechanism on the outside of the flute to cover each hole. This can be manipulated by the child's sound fingers (Fig. 4). The second type, for those who have more than one finger, such as a cleft hand, covers the original holes and opens new holes so that the

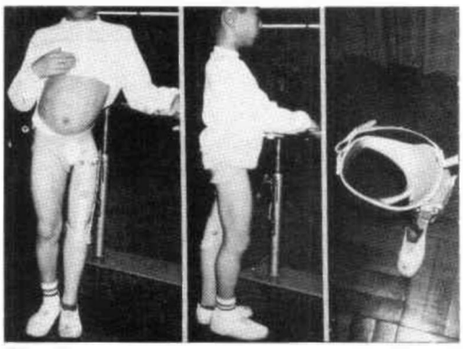


Fig. 2. Special socket design for unilateral hemipelvectomy (R. Takahashi).

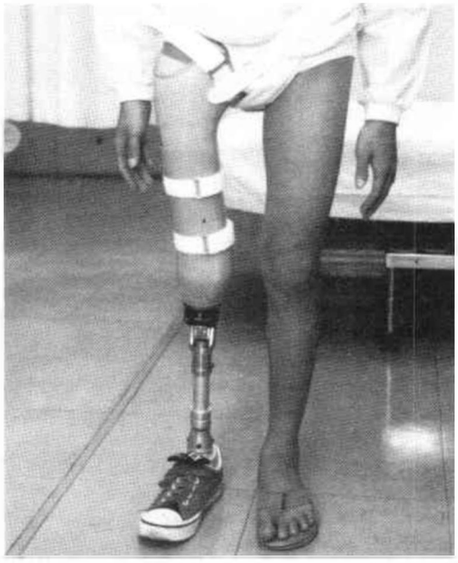


Fig. 3. Unilateral PFFD at two years old, fitted with extension brace and at 12 years old Syme's amputation was performed and the child was fitted with endoskeletal modular prosthesis with torque absorber made of titanium alloy.

Fig. 4. Special recorder-flute for the unilateral amputated child, the amputated side is fitted with Swedish myoelectric hand.

remaining digit can operate it quite easily. Since 1975, thanks to the special musical instruments, several thousand children have been able to play the recorder-flute with their peers.

Although the medical and social welfare problems of geriatrics have been overwhelming in recent years in this country, more effort should be continuously directed towards the welfare of limb deficient children.

REFERENCES

Betto, A., Kakurai, S., Asakura, K., Tazawa, Y. (1989). Development and clinical application of titanium child prosthetics with torque absorber. ISPO VI World Congress, 12th-17th November 1989, Kobe, Japan. p66,

Kakurai, S., Kakayam, T. (1983). Development of a recorder flute for unilateral upper limb amputated children. ISPO IV World Congress, 5th-9th September 1983, London, UK.

Kakarai, S., Aoyama, T., Nakajima, S., Satoh, M., Satoh, Y. (1989). Development of endoskeletal passive hand for the unilateral upper limb amputee. ISPO VI World Congress, 12th-17th November 1989, Kobe, Japan. p186.

Kida, M. (1987). Ed.: Thalidomide embryopathy in Japan. Tokyo: Kodansha.

Kida, M. (1989). The children with congenital malformation of the limbs and association of their parents. ISPO VI World Congress, 12th–17th November 1989, Kobe, Japan. p186.

Prosthetics and Orthotics International, 1991, 15, 152–155

The care of the limb deficient child in North Africa

N. SLIMAN, A. MRABET, S. DAGHFOUS and M. DOUIK

Institute El Kassab, Manouba, Tunisia

Abstract
The surgical and prosthetic treatment of longitudinal lower limb deficiency is described and discussed, in the light of cultural and social requirements. Those with upper limb deficiencies are not fitted with prostheses.

Introduction

Cases of congenital limb deficiency have always been a drama for the family, a handicap for the patient and a heavy burden for the society. The Kassab Institute has devoted much attention to these cases which are seen as being very important.

Our therapeutic attitude, which was initially conservative (due to the social and cultural context and to fitting difficulties) is becoming less restricted; the main objective of treatment is to allow the child to carry out the activities of his age group.

The purpose of this paper is to report our findings concerning the care and the course of the treatment of children suffering mainly from a failure of formation of the lower limb.

During the period between 1970–1990, 74 children (40 boys and 34 girls) received treatment at the Tunis orthopaedic Institute M. T. Kassab.

There were 88 affected limbs, 77 longitudinal deficiencies and 11 transverse deficiencies (12 patients with 16 associated upper limb anomalies).

Fibular deficiency (2 groups)

First group

Some 18 children (21 limbs) were seen at the Institute at an average age of six months (varying from 11 days to 11 months).

All correspondence to be addressed to Dr. N. Sliman, Institut d'Orthopédie M. T. Kassab, 2010 Manouba, Tunisia.

The surgical treatment was carried out at the age of 12 months in the following way:

— 14 patients had a foot correction and realignment for marked talipes equino-valgus (resection of fibrous lateral band, lengthening of achilles and peroneal tendons and a posterior capsulotomy).

— 8 had an osteotomy of the tibia for anterior bowing of this bone. After this surgery, walking was possible using an orthosis which contained the corrected foot and compensated for the shortening (Fig. 1).

During the course of the treatment, there was an increase of the shortening and the orthosis was given up by 11 who preferred walking on their toes without an orthosis. The equinus compensates for the shortening and the foot remains in axial alignment.

In three cases the equinus of the foot could not compensate for the shortening and a lengthening of the tibia by the Wagner procedure was performed. Four children were fitted with a definitive extension or ortho-prosthesis because of severe knee instability.

Second group

Fourteen older children were first seen between the ages of three and 16 years. Nine of them walk on their toes with moderate talipes

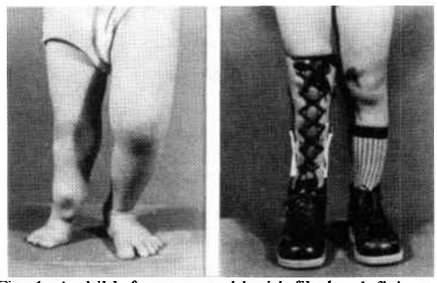


Fig. 1. A child, four years old with fibular deficiency, operated on and fitted with simple orthosis.

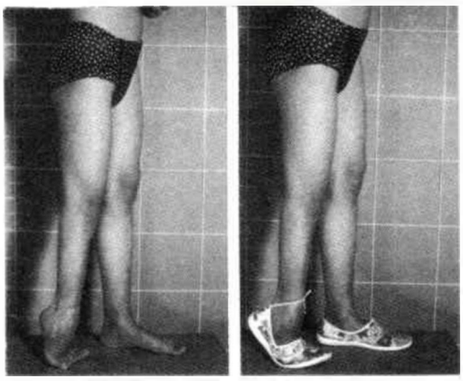


Fig. 2. A girl, 15 years old with fibular deficiency, walking without orthosis in a digitgrade way.

equino-valgus. They were treated conservatively as their walk was balanced, the knee stable and the shortening compensated by the equinus (Fig. 2). This approach is always accepted well by the parents.

Five walk with a pronounced limp due to shortening, knee instability, and in two cases hip dislocation. An ortho-prosthesis was fitted to four straight away and to the fifth after failure of surgical equalisation.

Tibial deficiency

Total absence — Type 1 (Table 1)

There were 10 patients who presented with a total of 14 limbs in this group.

Disarticulation of the knee followed by prosthetic fitting was always carried out at an average age of three years because of the severe knee flexion and foot deformity (Fig. 3).

Distal ⅔ partial absence — Type 2 (Table 1)

There were 10 patients who presented with a total of 11 limbs in this group.

Four patients had realignment surgery,

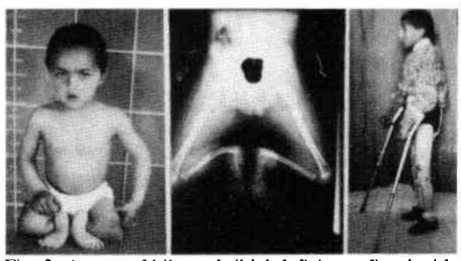


Fig. 3. A case of bilateral tibial deficiency fitted with prosthesis after knee disarticulation.

Table 1. Classification of tibial deficiency:
Type 1: total absence
Type 2: distal ⅔ partial absence
Type 3: distal ⅓ partial absence

Type 1	Type 2	Type 3
		
14	10	4

consisting of fixation of the foot under the fibula, followed by fusion of the fibula to the tibia when about two years old. An orthosis maintains the corrected alignment of the leg and foot and compensates for the shortening (Fig. 4). Three of these patients had leg lengthening later at the age of 10 using Wagner or Orthofix devices.

Six patients had severe foot deformity and marked fixed flexion of the knee. Despite all attempts to reconstruct, three had to have an amputation and three had knee disarticulation, followed by prosthetic fitting.

Distal ⅓ partial absence — Type 3 (Table 1)

There were 4 patients who presented with a total of 4 limbs in this group.

Table 2. Asmutz classification of proximal femoral deficiency

I	II	III	IV	V
				
6		4	3	1

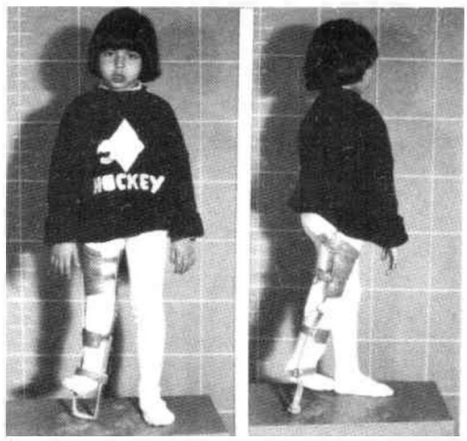


Fig. 4. A girl, five years old with tibial deficiency Type II, operated on and fitted with orthosis with compensation for shortening.

All presented with a stable knee and only minor foot deformity. All were fitted with an orthosis. Two have been lengthened and the others await this procedure.

Femoral deficiency

Amstutz – Type I (Table 2)

There were 6 patients who presented with a total of 6 limbs in this group.

Five patients underwent a correction of the coxa vara by valgus osteotomy of the femur. An extension or orthoprosthesis compensating for the marked shortening produced a balanced walk. In this group three children had a previous femoral equalisation at the age of 12 (femoral lengthening and contralateral epiphysiodesis).

Amstutz – Types III, IV and V (Table 2)

There were 5 patients who presented with a total of 8 limbs in this group.

These were treated conservatively with orthoprostheses because of the marked femoral hypoplasia (Fig. 5).

Transverse deficiency

Nine patients (11 limbs) were provided with prostheses whose type depended on the age, the level of loss and the social environment.

Results and discussion

Limb deformities are relatively rare in our country (less than 3 in 1,000 births), which corresponds to the classical data (Thevenin *et al.*, 1988; Schoenecker *et al.*, 1989; Briard and Boullegue, 1978). Our patients are independent despite the severity of the limb lesions and the simplicity of our therapeutic approach.

Ten are doing sedentary jobs, 10 are working in agriculture, 30 are still attending school and 11 could not find employment.

Fibular deficiency

No matter what degree of deformity and the age of the patient at first consultation, surgical intervention remains simple. Correction of the valgus is maintained and walking on the toes in equinus is well tolerated in our rural environment.

Tibial deficiency

The severe deformities associated with Type 1 cases and with those of Type 2 with severe foot deformity and knee laxity led to early amputation (Schoenecker *et al.*, 1989; Epps and Schneider, 1989; Frantz and O'Rahilly, 1961;

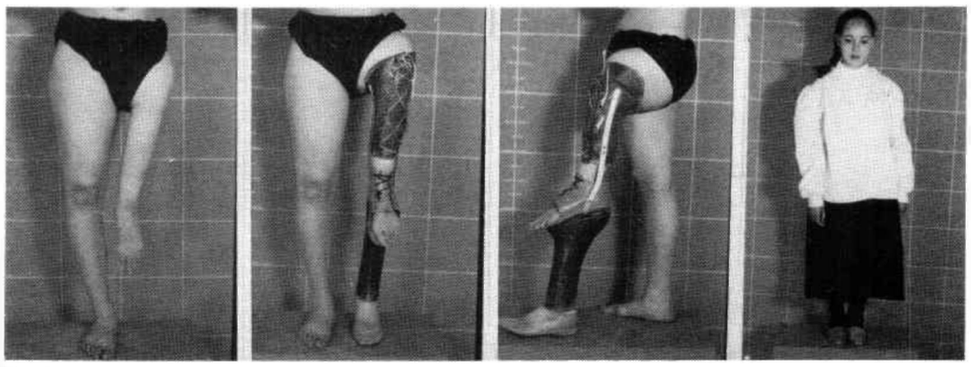


Fig. 5. A girl, 15 years old with partial femoral deficiency Type IV, fitted with ortho-prosthesis with which she is independent.

Bardot and Bouyala, 1978). Fitting bilateral cases remains difficult and the prostheses are not always tolerated, which reduces the patient's activity and heightens the psychological impact of amputation or disarticulation on the Tunisian patient personality.

Those with a stable knee whose foot can be preserved (some Type 2 and all Type 3) present a problem of length inequality. Contralateral tibial epiphysiodesis may avoid some complications.

Femoral deficiency

Correction of the coxa vara restores the architecture of the hip and provides femoral lengthening of 2–3 cm. The percentage of shortening remains constant compared to the length of the contralateral femur (Carlioz, 1978; Thevenin *et al.*, 1988). Final equalisation is performed at the age of 12 to 13 years to avoid multiple surgery. Rotationplasty has not been performed as it is always refused by the parents despite hip instability, malrotation with muscular inadequacy and even complete absence of femur (Amstutz Type 1). In these cases an ortho-prosthesis is accepted as the final treatment. No amputation has been carried out in this group of patients. Four adults prefer to use a wheelchair.

Upper limb deficiency associated with lower limb deficiency

Twelve patients have 16 upper limb deficiencies. They adapt easily to their handicap and are not fitted with prostheses.

Conclusion

Despite the complexity of these deformities the results remain satisfactory, being influenced by the severity of the shortening in the unilateral cases, the position and stability of the foot, knee and hip and deformity of the segments. We think the foot and leg should be preserved, maintaining the equinus position allows the use of an orthosis which is distal to the knee and allows better growth of the affected segment.

Amputation or disarticulation should only be used as an inferior solution in our social and economic environment.

REFERENCES

Bardot, A., Bouyala, J. M. (1978). Les grandes hypolasies et aplasies squellotiques congenitales des memores inférieurs chez l'enfant. Appareillage et chirurgie pere-prothétique. *Chir. Pédiatr.*, **19**, 361–380.

Briard, M. L., Boullegue, D. (1978). Etude familiale et epidemiologique des agénésies longitudinales des membres inférieurs. *Chir. Pédiatr.*, **19**, 301–305.

Carlioz, H. (1978). Description et histoire naturalle des grandes aplasies des membres inférieurs. *Chir. Pédiatr.*, **19**, 306–321.

Epps, C. H., Schneider, F. L. (1989). Treatment of hemimelia of the lower extremity. Longterm results. *J. Bone Joint Surg.*, **71A**, 273–277.

Frantz, C. H., O'Rahilly, R. (1961). Congenital skeletal limb deficiencies. *J. Bone Joint Surg.*, **43A**, 1202–1224.

Schoenecker, P. L., Cappelli, A. M., Miller, E. A., Sheen, M. R., Hamer, T., Aiona, M. D., Meyer, L. C. (1989). Congenital longitudinal deficiency of the tibia. *J. Bone Joint Surg.*, **71A**, 278–287.

Thevenin, D., Pilliard, D., Taussig, G. (1988). 154 agénésies des membres inférieurs chez l'enfant. *Rev. Chir. Orthop.*, **74**, suppl. 2, 109–111.

Prosthetics and Orthotics International, 1991, 15, 156–159

The care of the limb deficient child in Venezuela

F. FERNANDEZ-PALAZZI, D. P. DE GUTIERREZ, and R. PALADINO

Hospital San Juan de Dios, Caracas, Venezuela

Abstract

An overview of the situation in Venezuela is made by sampling the cases of congenital limb deficiency treated at the Hospital San Juan de Dios in Caracas from 1961–1989. The major longitudinal deficiencies are analysed.

The socioeconomic situation of the patients is of most importance being the cause of final decisions in relation to surgery, the ordering of prosthesis and orthosis, supplied mainly by the Venezuelan Institute of Social Security (IVSS), the maintenance of these items and the possibility of travelling to the hospital for diagnosis, treatment and follow-up.

All these transform the situation from a medical problem into a socio-economic problem typical of a third world country.

Introduction

Patients suffering from limb deficiencies present at birth in Venezuela, come from all over the country, to be seen and treated in the capital city of Caracas, where there are three hospitals able to treat these types of deformities. Of these three hospitals, two are of private administration under private foundations. These two hospitals are: Hospital San Juan de Dios, owned by the catholic order of the Fatebenefratelli Hospitalary Order and the Hospital Ortopédico Infantil owned by a private foundation, the Venezuelan Foundation against Poliomyelitis. The other hospital, Municipal Children's Hospital José Maria de Los Rios, is owned by the Municipal Government of Caracas. These hospitals take care of almost 75% of all children in the country as specialized hospitals. Another similar children's hospital, San Rafael Hospital, is located in Maracaibo, the second city of the country, and also belongs to the Fatebenefratelli Order, but children with congenital deficiencies are referred to the Caracas Hospital.

The Instituto Venezolano del Seguro Social (IVSS) is the official national health institution which takes care of the special requirements of those covered even when they are treated elsewhere, or for conditions not treated by other hospitals.

All correspondence to be addressed to Dr. F. Fernandez-Palazzi, Apartado de Correos 66473, Plaza Las Américas, Caracas 1061 A, Venezuela.

Method

On reviewing patients treated in the Authors' Hospital it was found that the most common congenital limb deficiencies seen were proximal femoral focal deficiency accounting for 27% of cases, and radial longitudinal deficiency (27%) followed by the fibular longitudinal deficiency with 23% and other deformities amounting to 23%. The male sex predominated at 51%.

Most of the patients are seen before two years of age. Amputation if required is not accepted by the parents as a primary choice. They refer other types of limb saving treatment and leave amputation as a last recourse at an older age in case of failure of the treatment selected. Thus the tibial longitudinal deficiency was amputated in 51% of cases, the fibular longitudinal deficiency in 30%, both after other types of procedures were performed and the proximal femoral focal defect was amputated in 15% of cases. In the latter (PFFD) the rotation-plasty was always refused possibly for cultural reasons and a Syme's type of amputation to allow proper prosthetic fitting was performed when required. The age for amputation was delayed until the child could stand or ambulate and very seldom performed as a primary procedure and then only in tibial longitudinal deficiencies where salvage of the limb was

Table 1. Congenital limb deficiencies presented at Hospital San Juan de Dios, 1961–1989

Deficiencies	Number of Patients	Number of Limbs
Proximal femoral focal deficiency	26	26
Radial longitudinal deficiency	26 (11 bilateral)	37
Fibular longitudinal deficiency	22 (2 bilateral)	24
Tibial longitudinal deficiency	17 (3 bilateral)	20
Tibial and fibular shortening	6	6
Total	97	113

recognised by the parents as "very difficult". The cost of the prosthesis or the orthosis required was covered mainly by the IVSS and less frequently by voluntary donations. The social status of the children, according to Graffar was "low". The maintenance and repair of the prosthesis and orthosis was carried out on a very irregular basis due to low financial income, living in places separated from the capital or main cities, low cultural level and poor understanding as to how to care for the items. Patients usually only come to the hospital when they require a new prosthesis or orthosis, because of total destruction or change for a larger size.

Material

A retrospective study was made on the records of patients seen at Hospital San Juan de Dios from 1961 to 1989 with a diagnosis of congenital deficient limb. It is to be stressed that surgery in this hospital is either free of charge, if the patient status requires it or according to different rates, depending on the patient's financial position, but always much cheaper than the cost of the same procedure in a private clinic. The IVSS covers the cost of those patients operated at their institutions under their affiliation. Other costs are covered by funds of private institutions or charity organisations.

In each case the authors determined:

1. type of congenital anomaly;
2. affected limb;
3. age when first seen;
4. sex;
5. social class (Graffar);
6. amputations, time and level;
7. orthosis or prosthesis and evolution.

A total of 97 patients were diagnosed with a congenital deficient limb. The deficiencies presented are shown in Table 1.

Proximal femoral focal deficiency

A total of 26 patients were seen, 14 left and 12 right, 14 were males and 12 females.

Age when first seen, between one month and 11 years with a mean of 2.5 years.

Social Class: 20 "low" and 6 "medium".

Amputation: Rotationplasty was always refused. All cases had a stabilization of the hip and one had a through-knee amputation and 5 a Syme's. The rest refused and wore special custom built prostheses.

The compensatory special orthoses and prostheses were obtained mainly by the IVSS (16 cases), by private donations in 8 cases and acquired by the patient or the family itself only in 2 cases. The maintenance of the prosthesis or orthosis is mostly irregular, generally the patient or family repair the item with easily obtainable hardware, only coming to the hospital for a new one.

Radial longitudinal deficiency

Total patients seen 26, with 37 affected hands. Age when first seen ranged from two months to 12 years. Once the deformity is corrected with gentle manipulation and a plaster of Paris cast, the correction of the deformity is maintained with Orthoplast splints and physiotherapy and occupational therapy is started as soon as possible to improve musculature and increase function. Up to 1983, cases requiring surgery were operated according to Riordan procedure (Faldini, 1957) but since 1984 early cutaneous Z-plasty at an early age is being done with better results (Celis and Fernandez-Palazzi, 1984).

Phocomelias

Most children refuse prostheses. Early teaching of foot use is established and thus the prostheses are used mainly by girls for cosmetic reasons.

Fibular longitudinal deficiency

Some 22 cases were seen, 9 left, 9 right and two bilateral. Age first seen ranged between 1 month and 16 years. All cases were "low class" (Graffar).

Above-knee amputation was performed in one case, below-knee in three cases and at Syme's level in three cases. The rest wore compensatory orthoses when required. Prostheses and orthoses are always supplied by IVSS.

Only half of the patients came regularly to follow-up because of other concomitant problem requiring consultation. The rest only came when a new prosthesis or orthosis was required.

Tibial longitudinal deficiency

A total of 17 patients were seen with 3 bilateral making a total of 20 cases, 10 legs were right and 10 left, 10 patients were boys and seven were girls. Age first seen ranged from one month to seven years.

Amputation level was in 8 cases through-knee, 3 cases had a Syme's amputation, 4 cases correction of the foot with talectomy (to allow for orthosis) and the rest refused treatment and disappeared from follow-up.

Concomitant shortening of tibia and fibula

A total of 6 patients presented, 4 boys and 2 girls. Age first seen ranged from two years to six years. All treated by compensatory orthosis supplied by IVSS.

Discussion

Taking the material seen at the Hospital San Juan de Dios as a statistically significant sample of the situation of the congenital deficient child in Venezuela some statements of importance can be made in conclusion.

Almost all patients belonged to the "low class" (70.73%), being mostly from rural areas. The long distance from the treatment centre is the most important cause of difficulty in the treatment, either from the impossibility of assistance to reach the special clinic because of the distance, or lack of money to cover the fare. Those living in big cities or the capital are also in the very low income population and are not able to assist or to buy the prosthesis or orthosis in cases not covered by the IVSS, and rely on charities or private donations.

When the prosthesis or orthosis needs to be repaired this is done by the patient or the family with materials obtained in normal hardware shops. Lower limb affected children had in 64% of cases a compensatory orthosis indicated and 36% a prosthesis. Of these the IVSS covered 60% of the prostheses or orthoses, 36% were obtained through private donations and only 4% were obtained directly by the family. The follow-up of the cases was also difficult and 10% of patients never came back after the first visit.

The above facts transform a medical problem into a social problem.

In the hospitals with special clinics for congenital deficient limbs and prosthetics and orthotics the treatment given to the patient is the most appropriate according to the social, cultural and economic limitations characteristic of the country, and allows the child to be reinstated to society and to lead a relatively normal life.

Note on situation in Argentina

G. M. Arendar and S. P. Aichenbaum
Buenos Aires, Argentina

The diagnosis and treatment of children with congenital deficiencies in Argentina is changing due to many factors, scientific advances, technological advances specifically in the medical world, equipment, rehabilitation, changes in socio-economic conditions and other factors typical of the rest of Latin America.

Historically in Argentina, institutions that took care of the treatment of congenital limb deficient children were private, non-profit making organizations such as ALPI (Asociación Para la Lucha Contra la Paralisis Infantil — Association Against Poliomyelitis) in Buenos Aires and CERENIL (Centro de Rehabilitación Para Ninos Lisiados — Rehabilitation Centre for Crippled Children) in Mar del Plata, which with its funds covered the necessary equipment required for comprehensive treatment. Institutions similar to the CERENIL were created in the principal cities of the country in the 1960's and a fellow was sent from this organization to be trained in design and construction of prostheses and orthoses in Chile and the U.S.A. so starting in 1966 a model pilot factory in Argentina.

The National Commission on Rehabilitation (Comisión Nacional de Rehabilitación) created

in 1960 wishing to systematise the treatment of all patients with disabilities of the locomotor systems created the school for prosthetics and orthotics that produced free of charge for the patient the prosthesis or orthosis required by him.

From 1970 the social organizations and systems of organized mutual aids, under pressure from the clients, started to provide orthoses and prostheses generally under agreements with private orthopaedic companies.

Nowadays patients with congenital malformations are treated in private or public institutions not separately. Lamentably there are only very few of such organizations able to give a comprehensive treatment similar to that given in more industrialised countries. Nevertheless the technical ability and capacity of the prosthetic technicians can be compared with anywhere in the world.

The Dirección Nacional de Rehabilitación Psicofisisca (National Direction of Psychophysical Rehabilitation), the Dirección Nacional Protección al Discapacitado (National Direction of Protection to the Disabled) and the Dirección Nacional de Emergencius Sociales (National Direction of Social Emergencies) are state owned organizations in charge of providing pecuniary aid and paying for prostheses and orthoses and special vehicles, and other aids for disabled patients who are without economic resources. The Municipal and Provincial Governments also assign human, physical and monetary resources for assistance and equipment to these patients in co-ordination with co-operative family organizations and beneficient institutions.

From the demographic point of view, after the increased incidence of these pathologies due to Thalidomide, nowadays limb deficiencies are considered non-frequent malformations and there are no official statistics of the incidence in Argentina.

REFERENCES

FALDINI, J. (1957). Manual de Ortopedia. — Buenos Aires: Editorial Inpress Generales., p123–125.

CELIS, L., FERNANDEZ-PALAZZI, F. (1984). Mano zamba radial congénita. Revista de la Sociedad Médica del Hospital San Juan de Dios, **5**, 97–107.

Prosthetics and Orthotics International, 1991, 15, 160–161

The function and operation of a parent support association

JOHN BRUCE

'Reach', The Association for Children with Hand or Arm Deficiency, UK

Abstract
The use of a parent support organisation and its responsibilities and limitations are described. The difference between a parent support organisation and an association of limb deficient children is discussed.

A parent support association

When a child is born with a limb deficiency the parents are immediately faced with interminable questions. The obvious ones are related to the disability, but since the mother is already in direct contact with a medical team, which she may reasonably expect to be able to cope with these, medical questions are not necessarily, to her, the most pressing or bedevilling.

We 'lay-parents' are normally ill-prepared for this type of birth defect. We have been forewarned of all the major catastrophes which can occur but have never had explained to us that this form of 'minor mishap' is even possible.

Immediate concerns tend to polarise around the "why?", "why me/us?" . . . The answers and explanations are rather unsatisfactory in that "we don't know" is not easy to relate to!

Leaving the cocoon of the maternity hospital also breaks a link with the medical profession and this is when the feeling of isolation, of being the only one like this in the whole world, can become utterly overwhelming.
Now! . . . is when there must be a 'Reach'.

If a support group does nothing else it can produce very convincing evidence that there are others around who have been there! To find other families to share experiences with is a wonderful cloud dispersant!

This alone, I think, is justification for there being a parent support group, however, I believe, our responsibility goes much deeper.

As, inevitably, we have a very wide cross-section of parents, with an almost equally diverse range of limb deficient children in our contact group, we build up a comprehensive insight into all aspects of the problems addressed and the treatments administered across the spectrum of limb deficiency. This does not mean we are becoming medically competent or that we know the correct treatment for any specific disorder. It does mean that we are well placed to provide links between families; matching those whose children are similar and putting them in touch with each other. We are also in a position to provide information about the range of medical specialists who may be useful to them.

The support and encouragement we have to offer can be given in a variety of ways.

Families can be introduced to each other on an individual basis, which may foster friendship and mean that there can be a mutual approach to the solution of problems.

The Association can co-ordinate and encourage the setting up of local branches where families can get together from time to time as a group. This may on occasion be semi-formal; a meeting with guest speakers, visits by adult amputees, health professionals, or informal; 'tea parties' or outings of various sorts.

Other links come through the formal 'administrative' structure of the Society. These are the 'benefits' of belonging to a membership organization.

The publication of a regular newsletter gives a forum for the sharing of experiences. It will be enhanced if contributions are encouraged from professionals as well as parents and, of course, the children themselves.

Note that in this activity correspondents may

All correspondence to be addressed to John Bruce, General Secretary of Reach, 13 Park Terrace, Crimchard, Chard, Somerset TA20 1LA, UK.

express views and relate experiences which are not necessarily those which would be supported by the parent organization. Provided this distinction is made clear, any contention so generated can only be regarded as healthy.

The other activity which is usual in 'our' sort of organization is the publication of informative fact sheets. One has to be very careful in the preparation of this type of material. It is important that we do not allow individual preferences or bias to creep in; that we inform without trying to influence.

In any discussion it will not be long before some suggestions are made that support should be given to some aspect of research. This can be a very fraught area for 'amateur' associations such as ours, and is one where advice from relevant professionals should always be sought and welcomed. It is as well to be clear at the outset that 'research' can mean different things to different people:

The first proposal which is likely to arise (at least from parents!) is for research into the causes of congenital limb deficiency. It is arguable whether this is suitable for a small parent-support organization to invest limited resources. As congenital limb deficiency is comparatively rare, and moreover has a wide variety of forms, it seems unlikely that statistical surveys will prove fruitful. Perhaps co-operation with a professional 'scientist' who is looking for causes into some specific birth-defect-type (by facilitating access to relevant members) is the more effective way forward.

There are many other subjects in the broad area of research which will be suggested. Physical characteristics and the treatment of different types of malformed limbs will interest practitioners involved in associated disciplines. When some project, either in pure research or the development of hardware, is in need of financial support, then it behoves us, in our support group context, to seek expert guidance as to the ultimate prospective usefulness of the investigation and the likelihood of its achievement.

I think all that I have written above was said, rather more succinctly by our 'Information Officer' when she said:

> "As an Association we are appreciative of the complexities of the hand and arm and the skill of those practitioners who specialise in the care and treatment of the limb.
>
> We cannot express support for or against any individual or Centre.
>
> Each case being individual we cannot express opinion regarding relevant or irrelevant treatments nor influence these.
>
> We can collect data from all sources — publications, practitioners, and patients — and make available such information as is advisable to relevant persons and link like with like among our families where applicable.
>
> Any individual expressing an opinion based on *their* experience must ensure that it is clearly understood to be a *personal statement* and is not on behalf of the Association."

My own, final, predication would be to suggest that we should be very aware of the distinction between a 'parent support group' and an 'association for children with limb deficiency'.

I am not sure 'we' always make this distinction.

After all, parents need support; children have to grow up to cope with their disability.

Prosthetics and Orthotics International, 1991, 15, 162

A personal experience

DAVID BAILEY

Cumbria, UK.

Abstract

David Bailey, who is 14 years old, has a left transverse total forearm deficiency and bilateral longitudinal femur partial deficiencies. The latter fall into Torode and Gillespie's Group I, with marked femoral shortening. A wedge osteotomy was performed for his gross right coxa vara, which was successful, but an intractable fixed flexion deformity of the right knee led eventually to a knee disarticulation being performed.

My experience

I have been wearing artificial limbs for 12½ years. First I wore an arm where I put the stump of my left arm in a socket and it was strapped on over my right shoulder. (My right arm is in full working order, in other words normal). It was so long ago since I first wore one, but I looked at cine films the other night and I think I must have really enjoyed having one. I had the arm when I was 18 months old and then artificial legs at about 2½ years old. They were the type of legs which were like pylons, these were the only legs which were a pain. I had to put my own legs into big heavy leather sockets which my Mum had to tie up of course because I cannot tie laces, but these, I suppose were the first step in the right direction. Later I had legs where the front bit was hinged and I slid my legs in and they were strapped up with velcro. These legs were very different. I could walk better because they were much lighter.

After this the legs did not change for a few years but my artificial arm changed. I got an electric arm instead of a hook. This looked more real and could open and close when I flexed my stomach on which I had a belt which activated the arm. I got to like the fitters and found new friends in the waiting room of all places.

When I was 7 years old I had an operation to put a metal plate in my right hip because there was something wrong. They called it by its technical name which had about 20 syllables. After the operation I wore plaster of Paris like a suit. It went from level with my arm-pit to the tip of my right leg but on the left side it stopped at my hip level.

I got over this quite fast but after the operation my knee stiffened up and my right leg was nearly at a right angle. This unforseen difficulty meant that my left leg was fine but they had to remake my right artificial leg with a hole in the back where the bottom half of my own leg stuck out. This made it difficult to walk and in winter it was cold. After two years of this, when I was 9, I was asked if I minded if my leg was amputated from the knee down. I agreed to this because I knew that I would have a much better leg and it would probably improve my walking. I had it done and my improvement was good.

I had full movement of my stump and it felt fine. My right prosthesis changed to a much more complicated one. It had a carbon fibre pole up the middle of the lower half and a locking knee. My stump was in a socket at the top and coming from the socket was a belt which I strapped round my waist to keep it on. The left leg stayed the same. My legs are like this now.

I do not wear an arm any more. I went through a change of myoelectric arm when I had my leg off but then stopped completely. I wore a normal hook or stiff hand arm on and off but now I do not wear one at all and I doubt I ever will.

Artificial limbs changed my life from just hopping about on the floor to walking and looking more normal. I am eternally grateful to all the fitters, the doctors and all the others for the past 12½ years and especially my family for putting up with me.

Prosthetics and Orthotics International, 1991, 15, 163

Book Review

Clinical Aspects of Lower Extremity Prosthetics.
Canadian Association of Prosthetists and Orthotists, 1991.
ISBN 0-921832-02-8.
215pp. C$59.95

A valuable book for the purpose of training and education in prosthetics — for which it was intended — this is also very interesting reading for every professional involved in the care and rehabilitation of amputees.

The first of an intended series of books on prosthetic training it is divided into four major categories;

I Amputee Rehabilitation considerations;
II Trans-tibial Prosthetics;
III Syme's Prosthetics;
IV Partial Foot Prosthetics.

In each of these sections the text is provided by the health professional most intimately involved with the amputee during the described process, thus providing an in-depth, concise knowledge.

The first section, which deals with rehabilitation considerations addresses the issue of the psycho-social impact of amputation. A consideration too often over-looked or misunderstood. This section deals clearly and compassionately with the various emotional stages experienced by the amputee. Beginning with the initial reactions of "Shock", "Denial" and "Anger" it continues through to the final desired stage of "Acceptance". It gives realistic examples of the treatment for any appropriate interaction with the amputee during each of the various stages. With that information established the reader is then led through a succinct presentation of the "Team Approach", "Post-operative Care", "Biomechanical considerations of the Phases of Gait" followed by a section on "Pathological Gait Characteristics". It finishes with a section on "Physiotherapy and Gait Training". This first section of the book is necessary information for prosthetic students. Interesting reading for associated professionals and beneficial insight information for family members and friends of recent amputees.

Sections Two and Three present the clinical and practical approaches to below-knee and Syme's level prosthetics, respectively. Beginning with surgical indications and procedures and continuing step-by-step through cast and measurement, cast modification, alignment, and temporary fitting techniques of various prosthetic designs.

Additionally, it outlines biomechanical considerations of socket fit and alignment and addresses the issue of problem-solving. Each phase is well complemented by a series of clear, clean technical drawings.

In the fourth and final section. Partial foot Prosthetics, the reader may be disappointed by a lack of content. While the format remains the same as for below-knee and Syme's level prosthetics, the entire section is but ten pages long and describes the four basic levels of partial foot amputations, casting procedures, cast modification and (very) basic prosthetic designs. While the quality of the information provided is good, the quantity of variable designs and up-dated materials is lacking in this chapter.

In a total and final assessment of this book I can say that every school of prosthetics would be well advised to use this book as a teachng tool and any prosthetic facility interested in a multi-faceted resource information for staff members and associated professionals will find it beneficial.

Dipl.—Ing. Wieland Kaphingst
Director
Federal School of Orthopaedic Technology
Dortmund, Germany

Proposed amendment to the Constitution

The following amendment to ISPO's Constitution has been formulated by the Committee of the UK National Member Society:

The International Constitution (4.2.2) limits the period of service for any person on the International Committee to a maximum of six consecutive years.

The UK NMS believes that this limit is undesirable because it is likely that anyone serving on the International Committee will take at least one three-year term before they are sufficiently familiar with the work to contribute effectively to the business. The present rule limits their effective contribution to the second three-year period or less.

In the UK NMS it is the practice to appoint a National Committee member to be the International Committee representative. The membership at large elects the whole National Committee every three years (five per year) and the National Committee itself selects the International Committee representative, reviewing the appointment every three years. There is, therefore, a fully democratic and repetitive two-tier process for selection of the International Committee representative.

Before the International Committee discusses this proposal, the Constitution requires that it be published to the International Committee and Members and Fellows for comment. Any such comment should be received by the Honorary Secretary before 1st February, 1992.

Original Clause	Proposed Clause
4.2.2 Persons on the International Committee will each have a 3-year term being eligible for re-election but may not serve more than six consecutive years with members taking office only at the conclusion of a Triennial Assembly. The International Committee will meet at least once every three years just prior to the Assembly meeting. The President may call an additional meeting of the International Committee at his discretion.	**4.2.2** Persons on the International Committee will each have a 3-year term being eligible for re-election with members taking office only at the conclusion of a Triennial Assembly. The International Committee will meet at least once every three years just prior to the Assembly meeting. The President may call an additional meeting of the International Committee at his discretion.

The Society is happy to announce the formation of a National Member Society in Finland.

Following is a list of the Office Bearers:

Chairman
Mr. L. B. Nummelin,
Vanha Topanilantie 73C 10,
SF-00730 Helsinki,
FINLAND

Vice Chairman
Mr. M. Kärkkäinen,
Tenholantie 12,
00280 Helsinki,
FINLAND

Secretary
Mr. T. Pohjolainen,
Raatepolkv 24,
02970 Espo,
FINLAND

International Newsletter
Summer 1991

United Kingdom National Member Society is making the final plans for Dundee '91, an International Conference on Orthotics, to be held in Dundee, Scotland, September 16–20, 1991. The conference is organized by the Department of Orthopaedic and Trauma Surgery, University of Dundee and the Tayside Rehabilitation Engineering Service, Dundee Limb Fitting Centre, Tayside Health Board. In addition to ISPO, the Association of Prosthetists and Orthotists, the Biological Engineering Society, and the British Orthopaedic Association, are sponsoring the event. David Condie, Secretary General of the Conference Committee, is assisted by Scientific Programme Organizers Amar Jain, David Rowley, and Michael Turner, and Exhibition Organizers John Lamb and Margaret Aitken. Dr. Colin Stewart is the Social Events Organizer and Jean Whyte directs the Secretariat. The scientific programme will examine the state of the art practices in rehabilitation through a series of invited presentations from an international faculty of experts, including Andre Bahler (Switerzerland), Bruce Baker (USA), Bill Doig (Australia), Thorkild Engen (USA), Hubert Etuern (Sweden), Acke Jernberger (Sweden), Michael Keith (USA), Alojz Kralj (Yugoslavia), Maude Malick (USA), John Michael (USA), Wallace Motloch (USA), Zaliha Omar (Malaysia), Michael Schuch (USA), P. K. Sethi (India), Robert Sorenson (Denmark), and Melvin Stills (USA) who will participate with a large faculty from the United Kingdom. Topics to be considered include material and manufacturing methods, functional electrical stimulation, disorders of the feet, joints and upper limbs, fractures, knee instability, spinal deformity, spinal cord injury, spina bifida, stroke, and cerebral palsy.

German National Member Society announces the newly elected board members, led by President R. Baumgartner, Vice-President G. Fitzlaff, Treasurer H. Gassenschmidt, 1st Secretary G. Welsch, and 2nd Secretary L. Bruckner. Board members are M. Budde, B. Peckmann, R. Forst, H. Perick, K. Fischer, and M. Schlafer.

Hong Kong National Member Society is planning the Fifth Joint Seminar with the Hong Kong Prosthetists and Orthotists Association. Speakers will include overseas experts and representatives from the Rehabilitation Engineering Centre of the Hong Kong Polytechnic. The Society has completed one volume on lower-limb prosthetics as part of a prosthetics and orthotics reference; Secretary-Treasurer Kwan Hung-hei edited the publication. John Lee Kwing-yue is Chairman of the Society.

United States National Member Society elected Michael Schuch Chairman at the recent meeting of the Executive Board. Bruce McClellan is Vice-Chairman, T. Walley Williams III is Secretary, and John Michael is the new Treasurer. Other Board members are Diane Atkins, Frank Gottschalk, Maurice LeBlanc, James Leonard, Rodney Pang, Terry Supan, and Ronald Spiers. Planning for the Seventh World Congress of ISPO in Chicago, Illinois in June 1992 is accelerating.

Australian National Member Society announced the Annual Scientific Meeting to be held in co-operation with the Department of Veterans' Affairs in Melbourne. The special guest speaker is Dudley Childress, Director of the Rehabilitation Engineering Program and Prosthetics Research Laboratory at Northwestern University Medical School. Dudley Childress is also General Secretary of the next ISPO World Congress. Melvin Stills, ISPO President-Elect, was the guest speaker at the Orthotics Seminar hosted by the Repatriation Artificial Limb and Appliance Centre in Sydney. He discussed orthotic treatment of spinal injuries and mangement of diabetic feet.

Joan G. Edelstein
Editor

Calendar of Events

National Centre for Training and Education in Prosthetics and Orthotics
Short Term Courses and Seminars 1991-92

Courses for Physicians, Surgeons and Therapists

NC503 Introductory Biomechanics; 7-11 October, 1991.
NC504 Lower Limb Orthotics; 25-29 November, 1991.
NC512 Orthotic Management of the Foot; 2-3 December, 1991.
NC505 Lower Limb Prosthetics; 3-7 February, 1992.
NC502 Upper Limb Prosthetics and Orthotics; 24–28 February, 1992.
NC510 Wheelchairs and Seating; 3–5 March, 1992
NC511 Clinical Gait Analysis; 11–13 March, 1992.
NC506 Fracture Bracing; 11–15 May, 1992.

Courses for Prosthetists

NC218A Ischial Containment Prosthetics; 14–18 October, 1991.
NC205 Above-Knee Prosthetics; 4–15 November, 1991.
NC211 PTB Prosthetics; 10–21 February, 1992.
NC218B Ischial Containment Prosthetics; 16–20 March, 1992.
NC212 Hip Disarticulation Prosthetics, 27 April–8 May, 1992.

Course for Orthotists and Therapists

NC217 Ankle-Foot Orthoses for the Management of the Cerebral Palsied Child; 1–3 April, 1992.

Course for Rehabilitation Engineers

NC801 CAD CAM; 19–21 November, 1991.

Seminars

NC718 Knee Orthotics; 1 November, 1991.
NC719 CAD CAM; 22 November, 1991.

Further information may be obtained by contacting Prof. J. Hughes, Director, National Centre for Training and Education in Prosthetics and Orthotics, University of Strathclyde, Curran Building, 131 St. James' Rd., Glasgow G4 0LS, Scotland. Tel: 041-552 4400 ext. 3298.

5–8 September, 1991
3rd Meeting of the European Academy of Childhood Disability, Manchester, England.
Information: Ms. O. Plunkett, MEIU, The Wolfson Centre, Mecklenburgh Square, London WC1N 2AP, England.

6–8 September, 1991
2nd Scientific Meeting of the Scandinavian Medical Society of Paraplegia, Copenhagen, Denmark.
Information: Centre for Spinal Cord Injured, Rigshospitalet, TH2002, Blegdamsvej 9, DK–2100 Copenhagen, Denmark.

16–20 September, 1991
Dundee '91 — International Conference and Instructional Course on Orthotics, Dundee, Scotland.
Information: Dundee '91 Secretariat, c/o Dundee Limb Fitting Centre, 133 Queen St., Broughty Ferry, Dundee, Scotland.

17–19 September, 1991
4th International Symposium on Biomedical Engineering, Peniscola, Spain.
Information: 4th International Symposium on Biomedical Engineering, Universidad Politecnica de Valencia, PO Box 22012, 46071–Valencia, Spain.

24–26 September, 1991
Biological Engineering Society Annual Scientific Meeting, Birmingham, England.
Information: Mrs. B. Freeman, BES, RCS, 35 Lincoln's Fields, London W2 3RX, England.

27–28 September, 1991
Biomedical Engineering Society, Annual Fall Meeting, Charlottesville, USA.
Information: Biomedical Engineering Society, PO Box 2399, Culver City, CA 90231, USA.

1–6 October, 1991
American Orthotic and Prosthetic Association Annual National Assembly, California, USA.
Information: AOPA, 717 Pendleton St., Alexandria, VA 22314, USA.

7–9 October, 1991
12th Annual Chinese Biomedical Electronics Conference, Shanghai, China.
Information: Prof. Tian-ge Zhung, Dept. of Biomedical Engineering, Shanghai Jiatong University, Shanghai 200030, China.

8–11 October, 1991
6th British Course on Knee Instability, Owestry, England.
Information: Erica Wilkinson, Symposium Secretary, The Robert Jones and Agnes Hunt Orthopaedic Hospital, Oswestry, Shropshire SY10 7AG, England.

9–12 October, 1991
45th Annual Meeting of the American Academy for Cerebral Palsy and Developmental Medicine, Kentucky, USA.
Information: AACPDM, PO Box 11086, Richmond, VA 23230-1086, USA.

11–12 October, 1991
Annual Scientifc Meeting of ISPO (Australia) and the Dept. of Veterans' Affairs, Melbourne, Australia.
Information: Convener, Central Development Unit, c/o Heidelberg Repatriation Hospital, Australia.

13–16 October, 1991
7th International Conference on Mechanics in Medicine and Biology, Ljubljana, Yugoslavia.
Information: ICMMB 91, Technical Organiser, CANKARJEV DOM, Cultural and Congress Centre, Kidricev Park 1, 61000 Ljubljana, Yugoslavia.

17–19 October, 1991
2nd Annual Meeting of the European Spine Society, Rome, Italy.
Information: Organising Secretariat, AISC, Viale Parioli 2,00197, Rome, Italy.

17–19 October, 1991
International Workshop on the Ergonomics of Manual Wheelchair Propulsion, Amsterdam, The Netherlands.
Information: Faculty of Human Movement Sciences, Van der Boechorststraat 9, 1081 BT Amsterdam, The Netherlands.

20–24 October, 1991
Western Orthopaedic Association Meeting, Tucson, USA.
Information: H. J. Martin, 2975 Treat Blvd., D–4, Concord, CA 94518, USA.

21–23 October, 1991
Combined Meeting of the Orthopaedic Research Societies of USA, Japan and Canada, Alberta, Canada.
Information: Mrs. M. Aldridge, Conference Office, University of Calgary, 2500 University Drive N.W., Calgary, Alberta T2N 1NR, Canada.

23–27 October, 1991
Reha '91–Rehabilitation Aids for Handicapped Persons, Dusseldorf, Germany.
Information: Reha '91 Press Office, Eva Rugenstein, Messe Dusseldorf, Germany.

27 October–1 November, 1991
American Academy of Physical Medicine and Rehabilitation Meeting, Washington DC, USA.
Information: AAPM&R, 78 E. Adams St., Suite 1300, Chicago, IL 60603, USA.

31 October–3 November, 1991
13th Annual Meeting of IEEE Engineering in Medicine and Biology Society, Orlando, USA.
Information: Professor Joachim Nagel, Dept. of Biomedical Engineering, Research Centre, University of Miami, PO Box 248294, Coral Gables, FL 33124, USA.

3–8 December, 1991
Annual Meeting of the American Academy of Neurological and Orthopaedic Surgeons, Las Vegas, USA.
Information: American Academy of Neurological and Orthopaedic Surgeons, 2320 Rancho Dr., Suite 108, Las Vegas, NV 89102, USA.

5–6 December, 1991
3rd International Symposium on Computer Simulation in Biomechanics, Perth, Australia.
Information: Ms. R. Ingham, Dept. of Human Movement Studies, Univ. of Western Australia, Nedlands WA 6009, Australia.

9–13 December, 1991
13th International Conference on Biomechanics, Perth, Australia
Information: 13th ISB Congress Secretariat, Dept. of Human Movement Studies, Univ. of Western Australia, Nedlands WA 6009, Australia.

1992

19–24 January, 1992
ACOPPRA II, the 2nd International Conference of the Central American Association of Orthotists, Prosthetists, Rehabilitation Professionals and Affiliates, Panama City, Panama, Central America.
Information: Rita Chan de Lee, Secretary ACCOPRA, PO Box 26, Zona 1, Panama City, Republic of Panama.

30 January–2 February, 1992
ISPO Course on Lower Limb Amputations and Related Prosthetics, Groningen, The Netherlands.
Information: ISPO Borgervaenget 15, 2100 Copenhagen Ø, Denmark.