BSSH The British Society for Surgery of the Hand

THE UNIVERSITY OF EDINBURGH

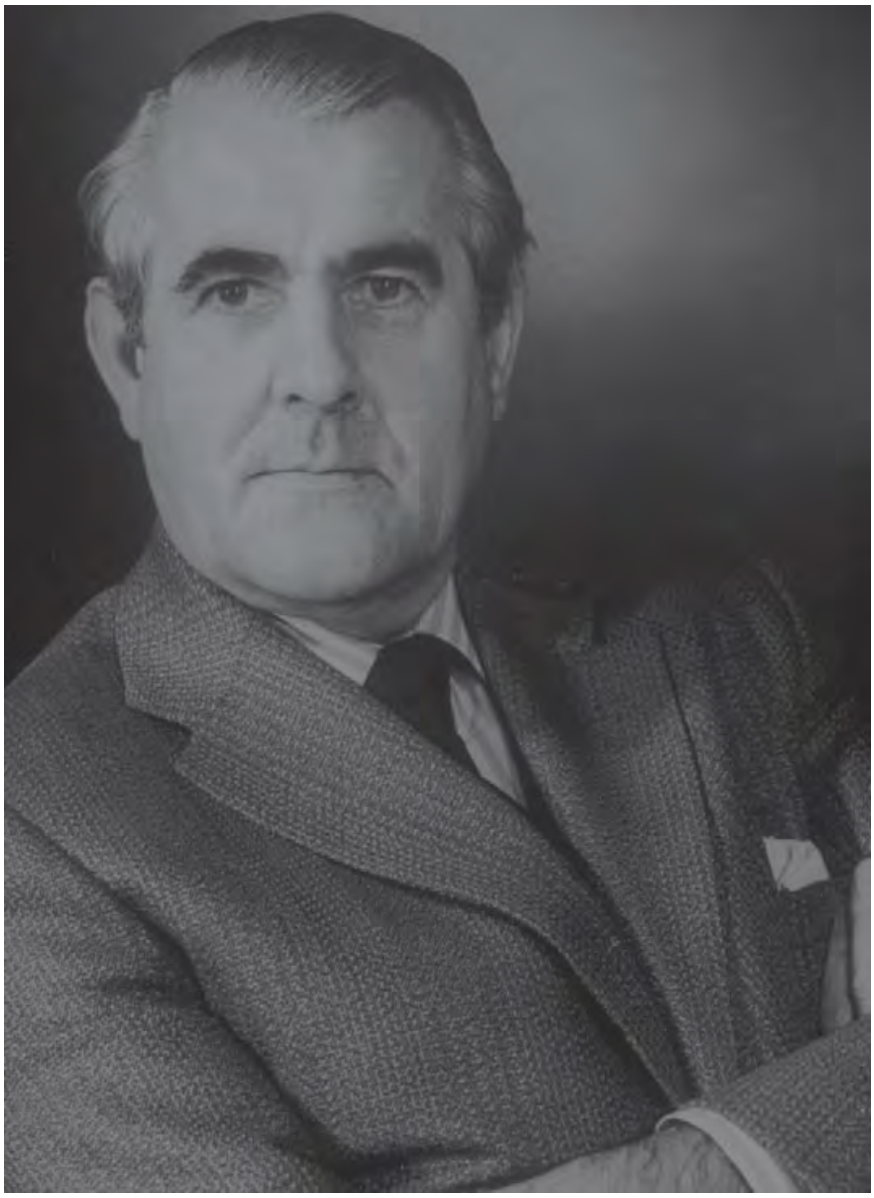
The Historical Archives of Douglas W. Lamb

Edited by Wee Leon Lam

Foreword by Geoffrey Hooper

EDITED BY: Wee Leon Lam

Edinburgh, UK



Douglas Watson Lamb

MBChB, FRCS Ed

Consultant orthopaedic and hand surgeon, Princess Margaret Rose Hospital, Edinburgh (1959-87)

Past President, International Federation of Societies for Surgery of the Hand (1989-1992)

Past President, British Society for Surgery of the Hand (1976-1977)

(b Edinburgh 1921; q Edinburgh 1944; FRCS Ed), d 11 December 2001.

Foreword

WHEN THE Thalidomide disaster occurred in the early 1960s, Douglas Lamb had the right qualifications to address the upper and lower limb problems that were encountered. He had extensive experience in amputation surgery and prosthetic fitting, and he had trained in the care of severely disabled patients at the Rancho Los Amigos National Rehabilitation Centre in Downey, California. The Princess Margaret Rose Orthopaedic Hospital ("PMROH") in Fairmilehead, Edinburgh, became the referral centre for affected children from Scotland. It was not long before Douglas's reputation for the care of these patients led to the referrals of children with many other types of congenital upper limb problems from around the United Kingdom and overseas. As a result of his experience, he was invited to speak all over the world about congenital anomalies of the upper limb, and he attracted national and international trainees who wished to gain from his knowledge of this formerly somewhat neglected subject. Mention is made elsewhere of his international contributions, but Douglas was particularly respected in the United States; he was an awarded the honorary fellowship of the American Society for Surgery of the Hand, a rare honour for a British surgeon.

I worked with Douglas for many years, from my first appointment as a trainee in orthopaedic surgery in Edinburgh in the 1970s to the time that I succeeded him in post in 1987. In person he was a physically large man, gentlemanly and dignified without being pompous. Although never intimidating, he would strike those new to his acquaintance as being somewhat serious, which indeed he was in his approach to clinical work. Like many Scots, he did not put his sense of humour on obvious display but his enjoyment of the amusing aspects of a particular situation was apparent to those who could read the signals. Those who worked with him knew that he valued their contributions and were therefore loyal to him. Although there were changes over the years, particular mention should be made of Helen Scott, the occupational therapist who dedicated her professional life to the care of children with congenital limb anomalies, Morag Wilson, his last secretary, and the late Mike Devlin, senior medical photographer at the PMROH, whose expertise is obvious in the material in this publication.

The clinical notes and images published in this book provide ample evidence of Douglas's approach to those with congenital upper limb problems. It is also interesting to see the conditions put in the context of current knowledge about the molecular basis for upper limb development and the Oberg-Manske-Tonkin classification. Wee Lam deserves particular thanks for the preservation of the original material. He and the British Society for Surgery of the Hand have done a great service in making this publication possible. I am sure that it will be read with interest.

Geoffrey Hooper *MMSc MD FRCSEng FRCSEd (Orth)*

Formerly Consultant Orthopaedic and Hand Surgeon, Princess Margaret Rose Orthopaedic Hospital, Edinburgh

North Queensferry, Fife

Acknowledgements

Many hours have been spent in the cataloguing, organising and arrangement of this book. Every one of those hours have been educational, informative, enjoyable and above all, inspiring. I would like to take this opportunity to thank Geoffrey Hooper, Pauline McGee, and Claudia Chan for helping me with the content. I would also like to thank the medical photography department, especially Amy Lumsden, André Syme, Mark Darling, Duncan Blyth and all the other staff at the Medical Photography Department in Edinburgh for their tireless efforts to see this book come to fruition.

Finally, I am grateful to the BSSH for supporting this project, especially Stephen Hodgson, BSSH honorary archivist and Jill Arrowsmith, past chair of the Educational and Training committee.

Contents

Introduction

THE BRITISH SOCIETY FOR SURGERY OF THE HAND (BSSH) hosts an inaugural lecture annually in honour of the late Douglas Lamb. The lectureship, entitled the 'Douglas Lamb Lecture' is awarded only to the most eminent of hand surgeons in recognition of their lifetime achievements. This eponymous lectureship is given in honour of the late Douglas Lamb, one of the most eminent hand surgeons from our society.

Douglas W. Lamb was an orthopaedic and hand surgeon at the Princess Margaret Rose Orthopaedic Hospital and the Royal Infirmary of Edinburgh. Nationally, he was instrumental in shaping the Second Hand Club, which later became known as the BSSH. He served faithfully as the Club's secretary for a long period of time, in the process helping to shape its future and making it become what it is today. He later became president of the BSSH in 1977 and ultimately reached what many would consider the summit of hand surgery: the Presidency of the International Federation of Societies for Surgery of the Hand (IFSSH).

Greatness can be defined in many ways. Douglas was considered by many to be great, not just in terms of his surgical brilliance but also for being the perfect gentleman. He trained many generations of hand surgeons and published numerous articles, several of which were first publications with junior hand surgeons, which then catapulted their careers.

Of all his academic pursuits, Douglas was perhaps most well-known for his work on congenital hand anomalies and paralytic disorders of the upper limb. His work on radial dysplasia have gone down in history as one of the cornerstones of treatment for this condition. To this day, his method of centralization is still being considered one of the gold standards for achieving long term results without compromising growth.

I first met Douglas when I was a medical student at the University of Edinburgh. At that time, I had the privilege of helping him collect data on the long-term follow-up results of centralization for patients with radial dysplasia, in preparation for a debate with Dieter Buck-Gramcko at an upcoming congenital hand conference in Paris. Many of the patients were survivors of the Thalidomide tragedy and were in their twenties and early thirties. It was an incredible experience to spend those months with Douglas and Helen Scott, his occupational therapist, and observing how they examined and talked to patients. To this day, I still remember those precious moments when he patiently taught a young medical student about the nuances of radial dysplasia, what he did for each patient in terms of tendon transfers and bony procedures, knowing perfectly well that I could not comprehend most of what he was telling me. To this day I wished I had a tape recorder, so I could record and playback what he said to me during the summer of 1995. Nonetheless, his patience and dedication to his patients left an indelible impression in me and made

me determined to pursue congenital hand surgery as my career of choice even at that young age.

In a way, this e-book represents an attempt to preserve some of the legacies of this great man. At the beginning of 2020, I was informed of a set of medical notes belonging to patients under his care that were about to be destroyed and whether I would be interested to take a look at them. What confronted me were nearly 450 sets of notes containing priceless photographs of patients, handwritten notes from Douglas to his patients, as well as from them to him, and operative details and clinic correspondences that he had dictated. I decided there and then that I would try my best not to let these notes be destroyed without preserving some of their contents, and that I will catalogue and organize them in a format that may be of benefit to future generations of hand surgeons. I also felt a sense of responsibility that this e-book, when completed, would allow others to know him in a way that I have had the privilege of doing so.

It is impossible to share information from all 450 notes. As several of them were congenital hand anomalies, I have made an attempt to classify some of them according to the current IFSSH Classification system, namely the Oberg, Manske, and Tonkin (OMT) system. I have followed this system through the various categories of malformations, deformation, dysplasia, and syndromes. Within each category, I have chosen representative cases with photographs and provide a brief commentary on what Douglas said and his thoughts on diagnosis and management. As far as possible, I have tried to include Douglas's own writings, after carefully anonymising all patient details. I have also included his own literature on the topic, where applicable. Finally, I have included my own thoughts on how things may have changed more recently, so as to stimulate the reader to further reading.

Wee Leon Lam *MBChB MPhil FRCS (Plast)*

Edinburgh,

December 2022

IA. MALFORMATION OF THE ENTIRE UPPER LIMB:

ABNORMAL AXIS FORMATION (EARLY LIMB PATTERNING)

IN THIS CATEGORY, we start with malformations affecting the entire upper limb (IA), which means that the pathology probably happened during early limb development, hence the term 'early limb patterning'. In many of these conditions, the entire upper limb is affected, i.e., the upper arm, forearm, as well as hand, in contrast to malformations affecting the hand plate only (late patterning, IB).

I-A-1. Malformation of the Entire Upper Limb:

Proximal-distal Axis

THE PROXIMAL-DISTAL AXIS controls limb development and growth from the embryo outwards until the digits are fully formed. The signalling centre has been established to be the apical ectodermal ridge with fibroblast growth factor (FGF) as the primary protein involved. Pathologies in this axis leads to the conditions of symbrachydactyly, transverse arrest and phocomelia.

I-A-1-ii. Symbrachydactyly

SYMBRACHYDACTYLY is a condition usually characterized by a short upper limb with 'nubbins'. Nubbins contain ectodermal elements (e.g., skin, nails) and represent an attempt by the developing limb to keep growing even after early truncation in the proximal distal axis. In the new OMT system, symbrachydactyly has been distinguished from transverse arrest, (see *Case 1*) which usually does not contain any distal ectodermal elements. It is important to check for a strong association of Poland syndrome in these patients, as characterised by an absent sternal part of the pectoralis major muscle.

CASE 1: *Symbrachydactyly*

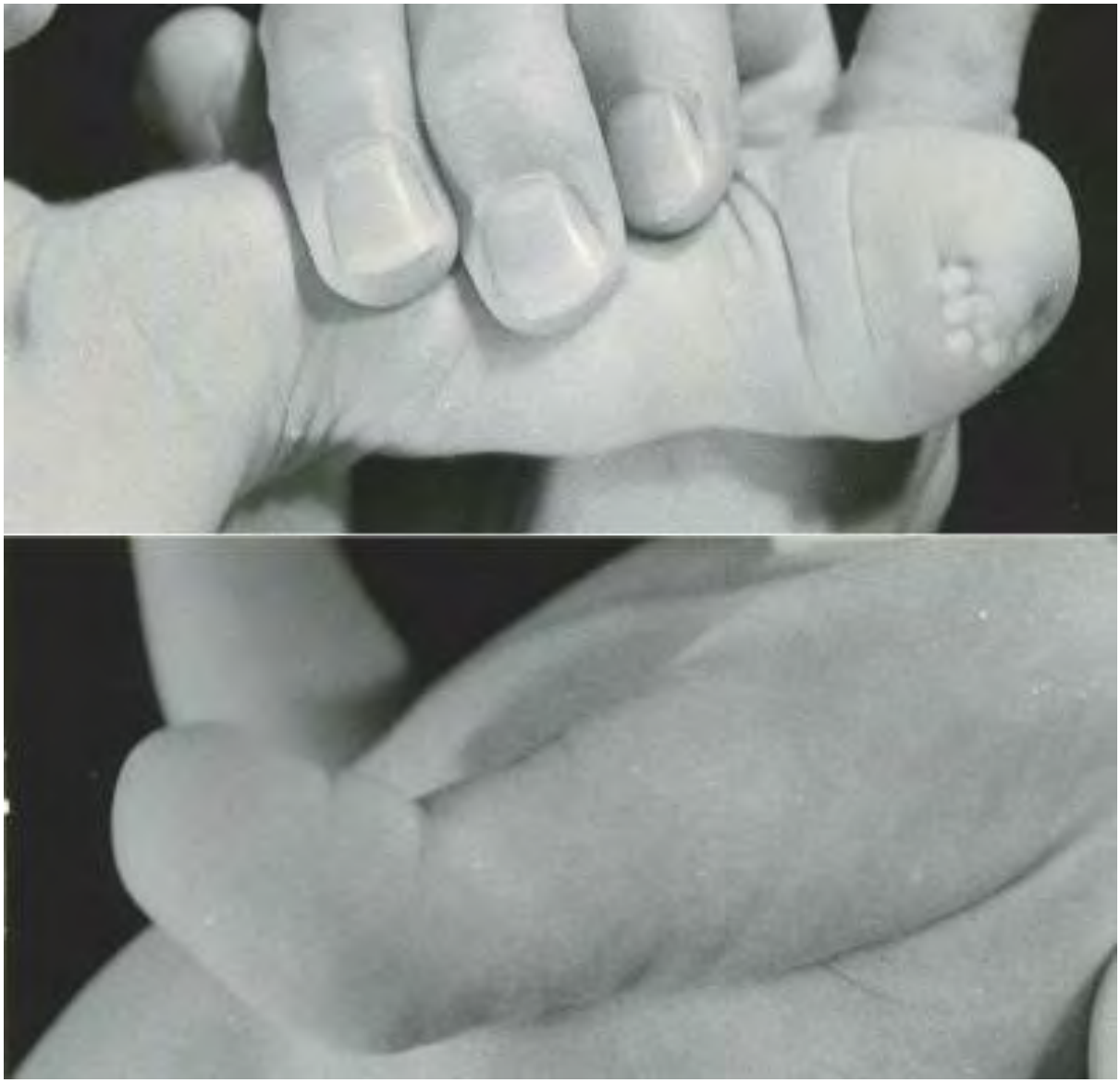


Figure 1: Symbrachydactyly of the left forearm with small digital nubbins. This patient has a short forearm with nubbins clearly present distally.

CASE 1 (continued): *Symbrachydactyly*

Mr D Lamb
Orthopaedic Surgeon
Princess Margaret Rose Hospital
Fairmilehead
Edinburgh Scotland

Dear Mr Lamb,

When Dad came back from Scotland he showed me the photos of the new type of arm you are inventing. It looks real and good. I hope that I will be able to have one, and help you in designing it. We arrive in Scotland on June the seventeenth. We will be living here until we find our own house. We leave Hamilton on the 28th or 29th of May. [redacted] I'm looking forward to seeing you again. I don't know if you will recognise me because I have grown a lot. I am almost ten years old.

Figure 2: A letter written by the patient to Douglas Lamb demonstrating the degree of affection patients had for him.

CASE 2: *Symbrachydactyly*

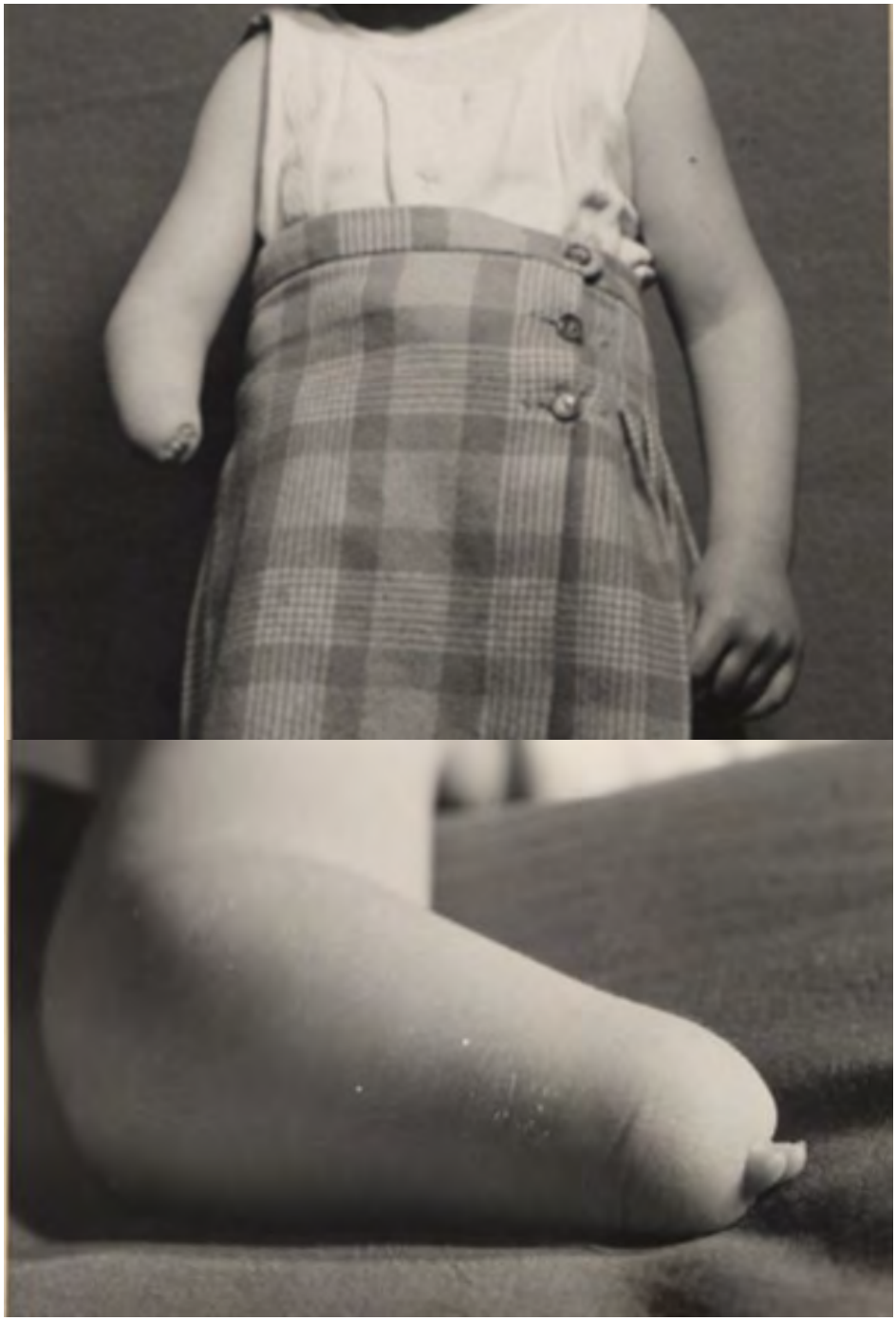
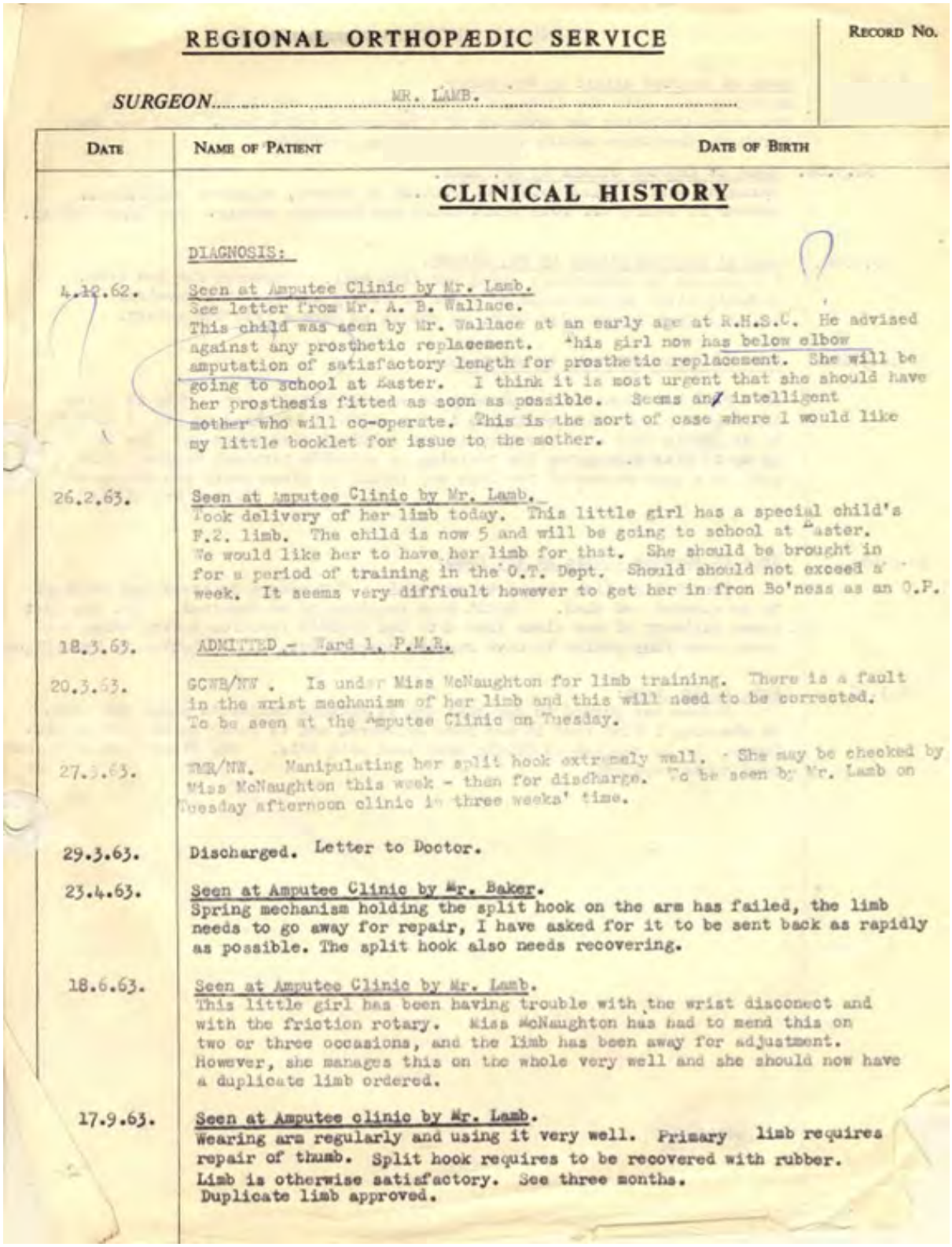


Figure 3 : Another patient with a short forearm, truncated at the mid forearm level with distal nubbins. Many of these patients were fitted with a prosthesis.

From the notes, Douglas would regularly see his patients and check on their prosthetic fitting himself.

CASE 2 (continued): *Symbrachydactyly*



REGIONAL ORTHOPÆDIC SERVICE

RECORD NO.

SURGEON..... MR. LAMB.

DATE	NAME OF PATIENT	DATE OF BIRTH

CLINICAL HISTORY

DIAGNOSIS:

4.12.62. Seen at Amputee Clinic by Mr. Lamb.
See letter from Mr. A. B. Wallace.
This child was seen by Mr. Wallace at an early age at R.H.S.C. He advised against any prosthetic replacement. This girl now has below elbow amputation of satisfactory length for prosthetic replacement. She will be going to school at Easter. I think it is most urgent that she should have her prosthesis fitted as soon as possible. Seems an intelligent mother who will co-operate. This is the sort of case where I would like my little booklet for issue to the mother.

26.2.63. Seen at Amputee Clinic by Mr. Lamb.
Took delivery of her limb today. This little girl has a special child's F.2. limb. The child is now 5 and will be going to school at Easter. We would like her to have her limb for that. She should be brought in for a period of training in the O.T. Dept. Should should not exceed a week. It seems very difficult however to get her in fron Bo'ness as an O.P.

18.3.63. ADMITTED - Ward 1, P.M.R.

20.3.63. GCWB/NW . Is under Miss McNaughton for limb training. There is a fault in the wrist mechanism of her limb and this will need to be corrected. To be seen at the Amputee Clinic on Tuesday.

27.3.63. WMR/NW. Manipulating her split hook extremely well. She may be checked by Miss McNaughton this week - then for discharge. To be seen by Mr. Lamb on Tuesday afternoon clinic in three weeks' time.

29.3.63. Discharged. Letter to Doctor.

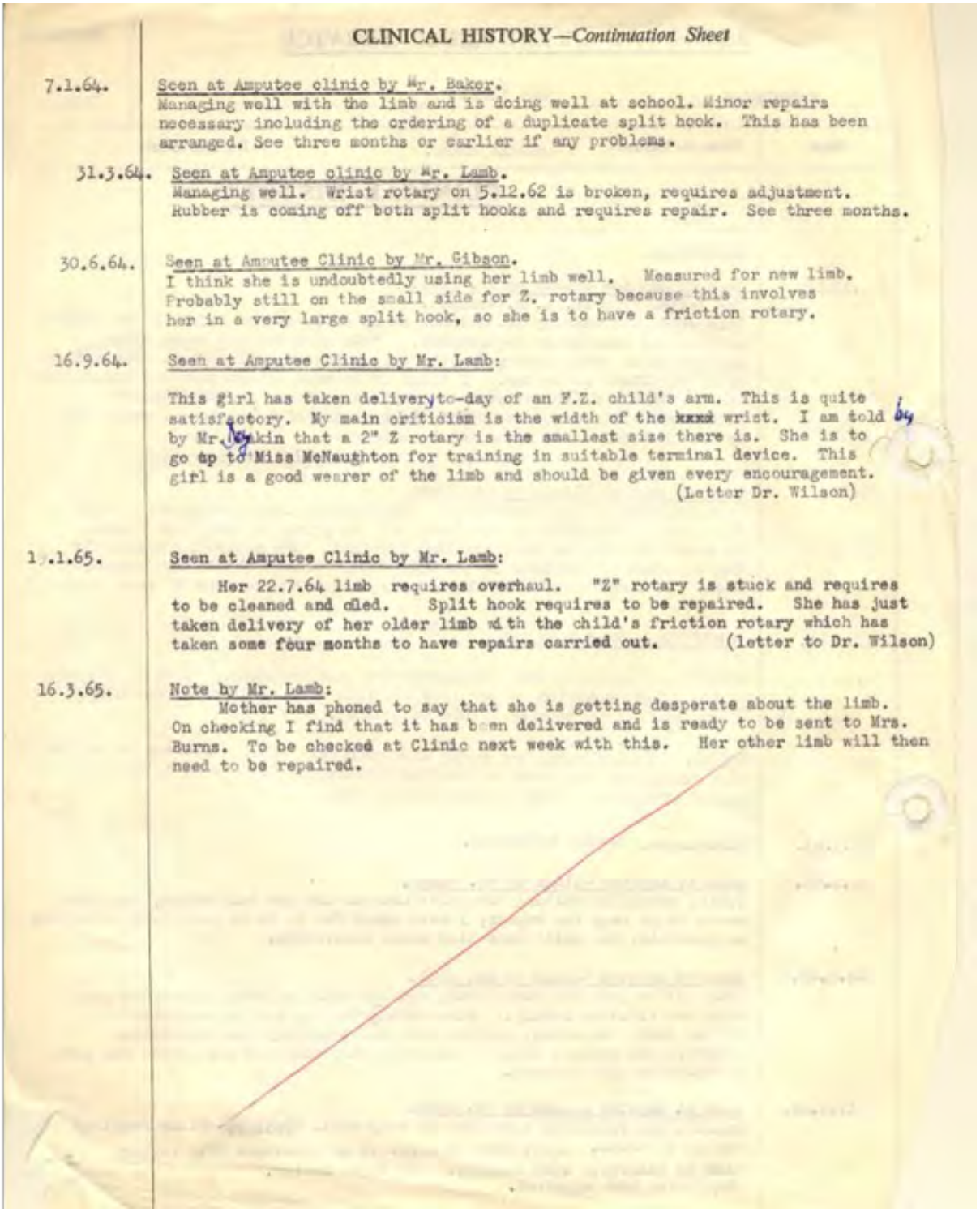
23.4.63. Seen at Amputee Clinic by Mr. Baker.
Spring mechanism holding the split hook on the arm has failed, the limb needs to go away for repair, I have asked for it to be sent back as rapidly as possible. The split hook also needs recovering.

18.6.63. Seen at Amputee Clinic by Mr. Lamb.
This little girl has been having trouble with the wrist disconect and with the friction rotary. Miss McNaughton has had to mend this on two or three occasions, and the limb has been away for adjustment. However, she manages this on the whole very well and she should now have a duplicate limb ordered.

17.9.63. Seen at Amputee clinic by Mr. Lamb.
Wearing arm regularly and using it very well. Primary limb requires repair of thumb. Split hook requires to be recovered with rubber. Limb is otherwise satisfactory. See three months.
Duplicate limb approved.

Figure 4: Clinical notes written by Douglas Lamb

CASE 2 (continued): *Symbrachydactyly*



CLINICAL HISTORY—*Continuation Sheet*

7.1.64. Seen at Amputee clinic by Mr. Baker.
Managing well with the limb and is doing well at school. Minor repairs necessary including the ordering of a duplicate split hook. This has been arranged. See three months or earlier if any problems.

31.3.64. Seen at Amputee clinic by Mr. Lamb.
Managing well. Wrist rotary on 5.12.62 is broken, requires adjustment. Rubber is coming off both split hooks and requires repair. See three months.

30.6.64. Seen at Amputee Clinic by Mr. Gibson.
I think she is undoubtedly using her limb well. Measured for new limb. Probably still on the small side for Z. rotary because this involves her in a very large split hook, so she is to have a friction rotary.

16.9.64. Seen at Amputee Clinic by Mr. Lamb:

This girl has taken delivery to-day of an F.Z. child's arm. This is quite satisfactory. My main criticism is the width of the xxxx wrist. I am told by Mr. Jenkin that a 2" Z rotary is the smallest size there is. She is to go up to Miss McNaughton for training in suitable terminal device. This girl is a good wearer of the limb and should be given every encouragement.
(Letter Dr. Wilson)

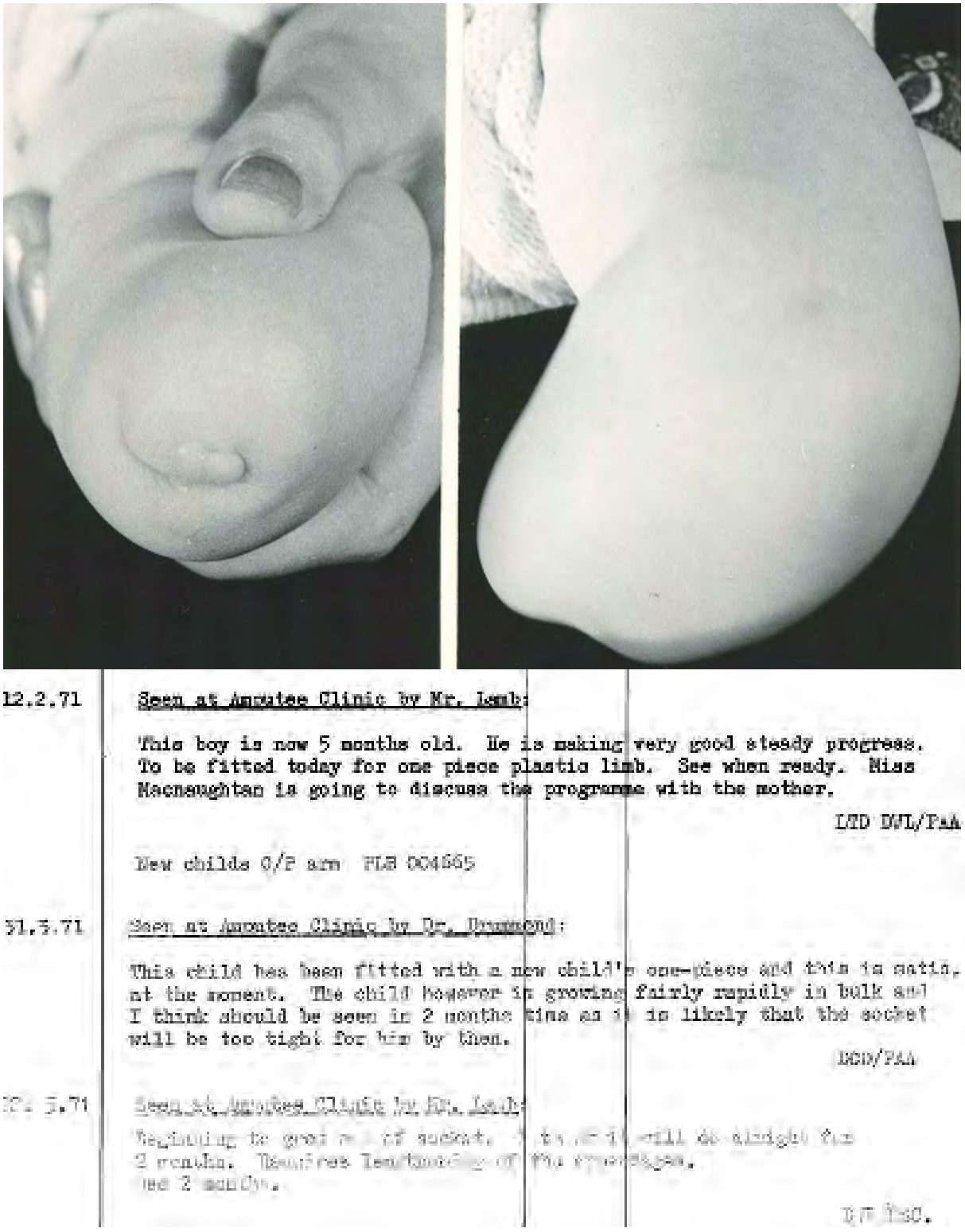
1.1.65. Seen at Amputee Clinic by Mr. Lamb:

Her 22.7.64 limb requires overhaul. "Z" rotary is stuck and requires to be cleaned and oiled. Split hook requires to be repaired. She has just taken delivery of her older limb with the child's friction rotary which has taken some four months to have repairs carried out. (letter to Dr. Wilson)

16.3.65. Note by Mr. Lamb:
Mother has phoned to say that she is getting desperate about the limb. On checking I find that it has been delivered and is ready to be sent to Mrs. Burns. To be checked at Clinic next week with this. Her other limb will then need to be repaired.

Figure 4 (continued): Clinical notes written by Douglas Lamb

CASE 3: *Symbrachydactyly*



12.2.71 Seen at Amputee Clinic by Mr. Lamb:

This boy is now 5 months old. He is making very good steady progress. To be fitted today for one piece plastic limb. See when ready. Miss Macnaughtan is going to discuss the programme with the mother.

LTD D/L/PAA

New childs O/P arm PLB 004965

31.3.71 Seen at Amputee Clinic by Dr. Drummond:

This child has been fitted with a new child's one-piece and this is satis. at the moment. The child however is growing fairly rapidly in bulk and I think should be seen in 2 months time as it is likely that the socket will be too tight for him by then.

DCD/PAA

[illegible] 3.71 Seen at Amputee Clinic by Mr. Lamb:

[illegible] to grow [illegible] of socket. [illegible] will do alright for 2 months. Requires lengthening of the [illegible] 2 months.

D/T Sec.

Figure 5: A patient with symbrachydactyly of the proximal forearm and clinical notes. In his practice, Douglas would regularly fit very young children with a prosthesis, sometimes as early as five months old.

CASE 4: *Symbrachydactyly*

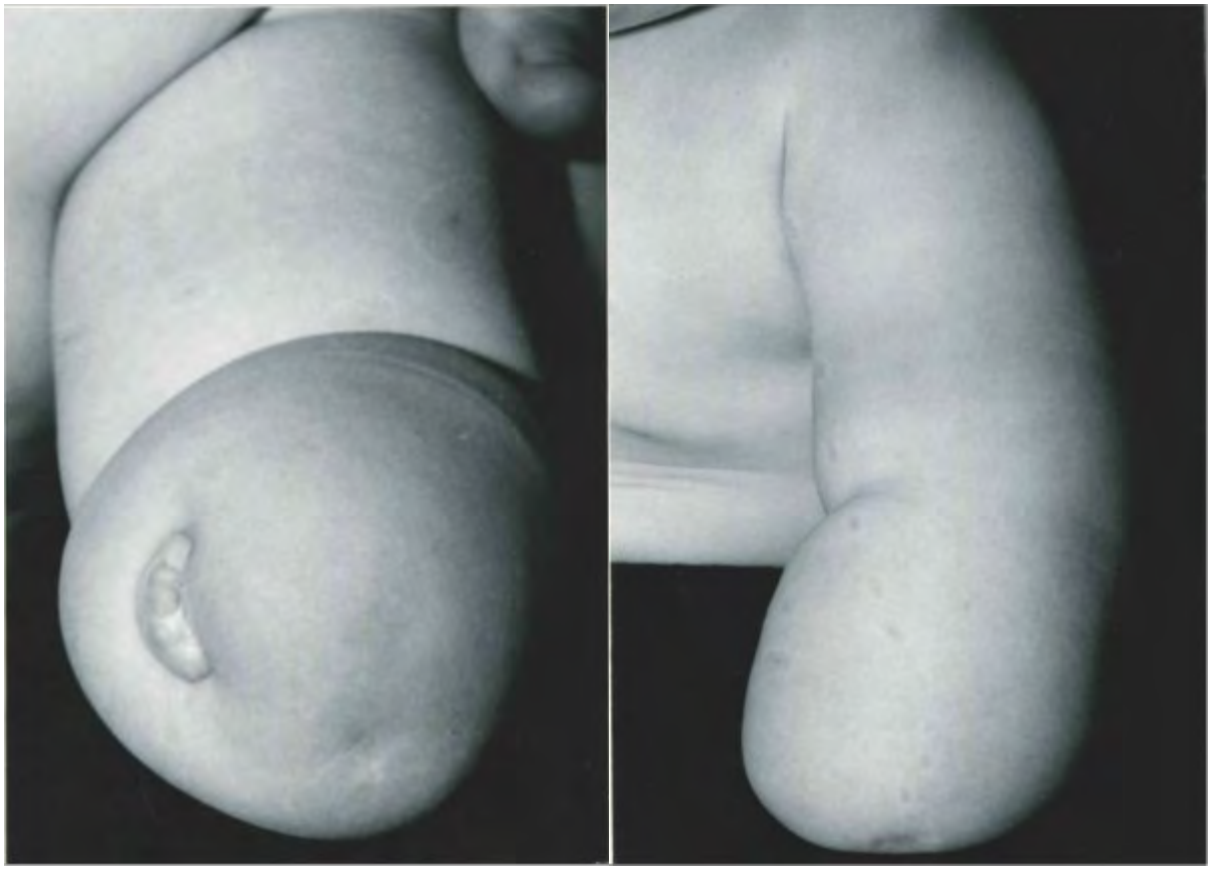


Figure 6: Symbrachydactyly of the left forearm. Again, distal dermal elements are seen, indicating this is a symbrachydactyly rather than a transverse arrest. The child has also been fitted with a prosthesis, including a split hook as well as myoelectric prosthesis.

I-A-1-iii. Transverse Deficiency

AS COMPARED TO SYMBRACHYDACTYLY, a transverse arrest is a congenital amputation of the upper limb without further evidence of distal ectodermal elements (nubbins). This implies there was a complete truncation of limb development in the proximal distal axis without any efforts for continued growth of the distal limb bud. Again, in his practice, it was quite evident that Douglas would advocate early fitting of upper limb prostheses in these patients.

CASE 5: *Transverse Deficiency*

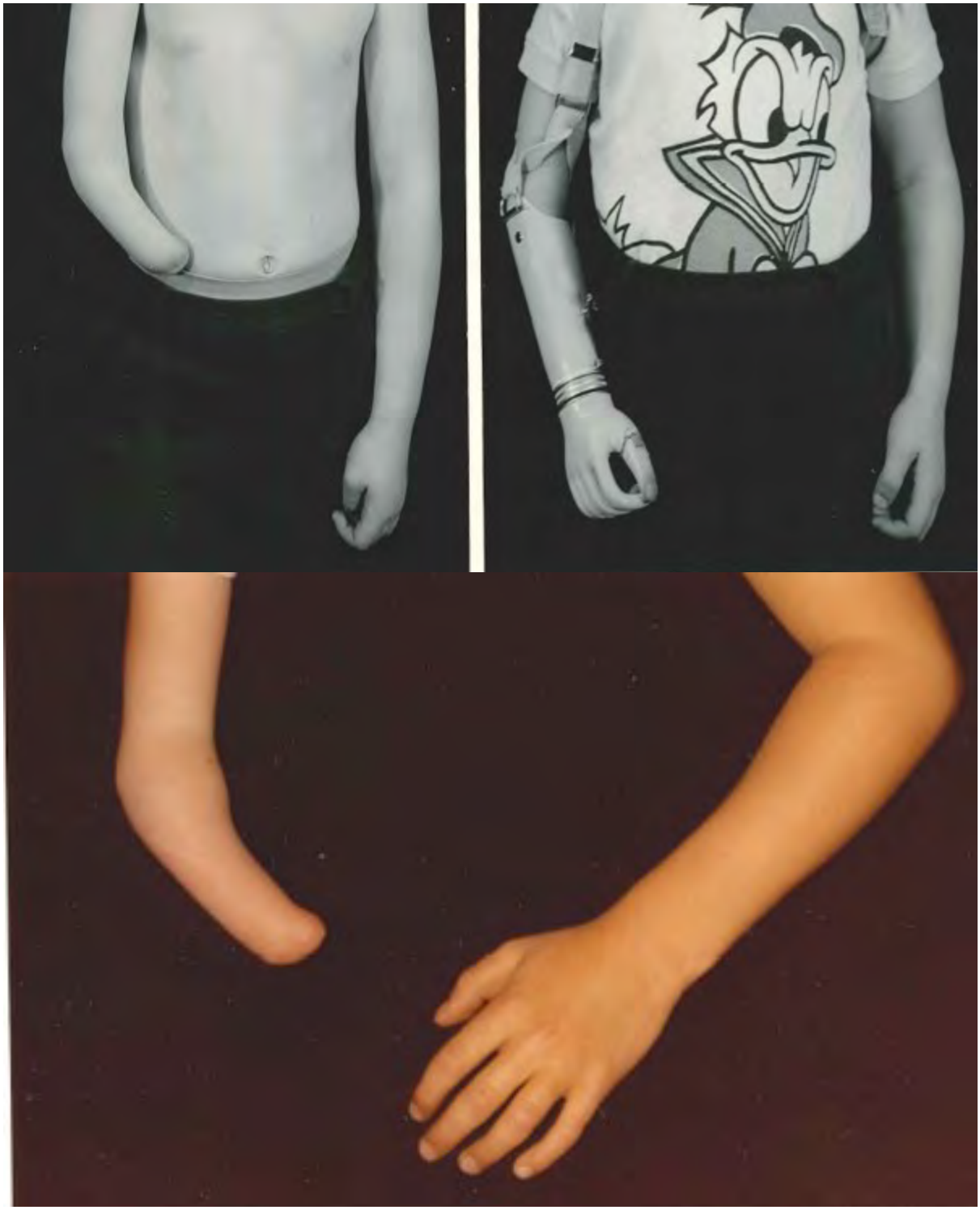


Figure 7: Transverse arrest at the proximal forearm level. The child has been fitted with a myoelectric prosthesis. From his notes, it is interesting to see how prosthetic fitting started when the child was only six months old. Children are seen quite regularly, mainly for any issues with the prosthesis. The focus was on socket revision, as they quickly outgrew these in the first few years of growth.

CASE 6: *Transverse Deficiency*

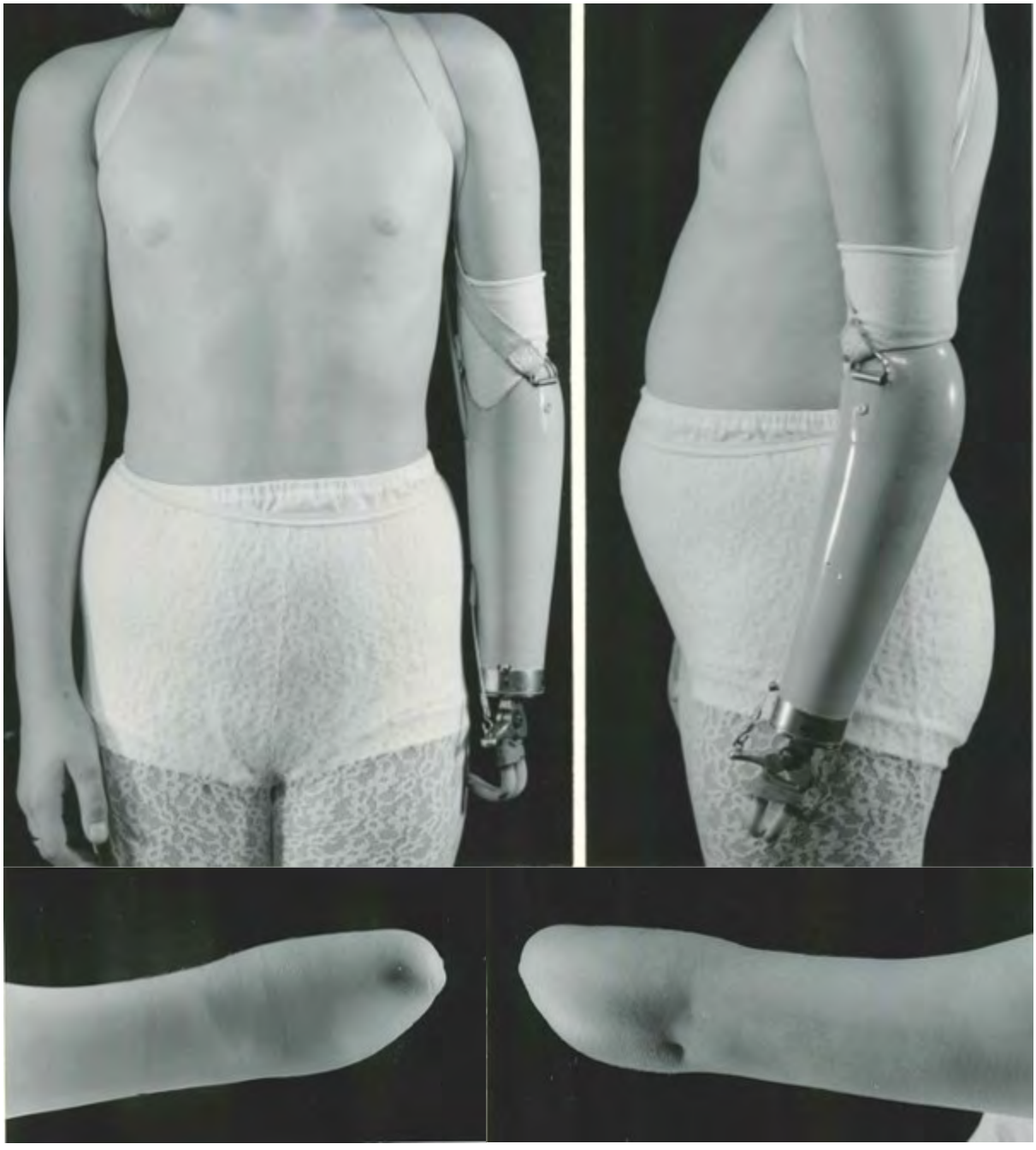


Figure 8: Left transverse arrest at the proximal forearm level. The child has been fitted with a split hook prosthesis. And again, the emphasis is on prosthesis function and incorporating this into activities of daily life.

I-A-1-iv. Intersegmental Deficiency (Phocomelia)

PHOCOMELIA is the next major subgroup in the OMT system, under malformations affecting the entire upper limbs in the proximal distal axis. Phocomelia is a term used to describe a deformity in which a limb usually resembles a 'Flipper-like' appendage. The hand or at least some of the digital components, are always present unlike transverse arrest. Frantz and O'Rahilly (1961) classified phocomelia into three groups: type 1 which is complete phocomelia or where the hand is directly attached to the trunk; type 2 is proximal phocomelia, where the humerus is missing and the forearm and hand are attached to the trunk; type 3 phocomelia is distal, where the hand is directly attached to the humerus due to absent forearm elements. The idea of phocomelia is that there is an 'intersegmental' deficiency, i.e., in-between segments are missing .

However, this classification system has been criticised several times, as in reality it is rare to find truly intersegmental conditions. The paper by Tytherleigh-Strong and Hooper (2003) found that only 11 out of 24 phocomelic limbs in their series could be classified using the system by Frantz and O'Rahilly. In this paper, they refuted the concept that phocomelia consisted of intercalary defects but represented perhaps more of a severe longitudinal deficiency.

This is a view shared by Goldfarb et al (2005), who further classified phocomelia into four different types: a proximal radial longitudinal dysplasia, a proximal ulnar longitudinal dysplasia and two types of severe combined dysplasias: type A characterised by absence of the forearm (humerus to hand or Frantz and O'Rahilly type 3 and type two, characterised by absence of the arm and forearm (hand to thorax or Frantz and O'Rahilly type 1).

In the archives, we found a myriad of terms used to describe 'phocomelic' conditions in patients including 'congenital absence', 'amputation', 'missing limb/hand', 'thalidomide deformities', or 'phocomelia'. This represents an evolution of nomenclature from the 1970s to present. Certainly, there is a need to revise the 2020 OMT classification as currently the division into proximal, distal, and proximal and distal deficiencies (according to Frantz and O'Rahilly's classification) may be too artificial. Several of the conditions we see probably belong more in the severe longitudinal dysplasia categories as described by Goldfarb et al (2005). For the purpose of this e-book, we have listed down the original classification by Lamb, and then the proposed classifications by Goldfarb et al (2005) and Frantz and O'Rahilly (1961), wherever possible.

CASE 7: *Phocomelia*

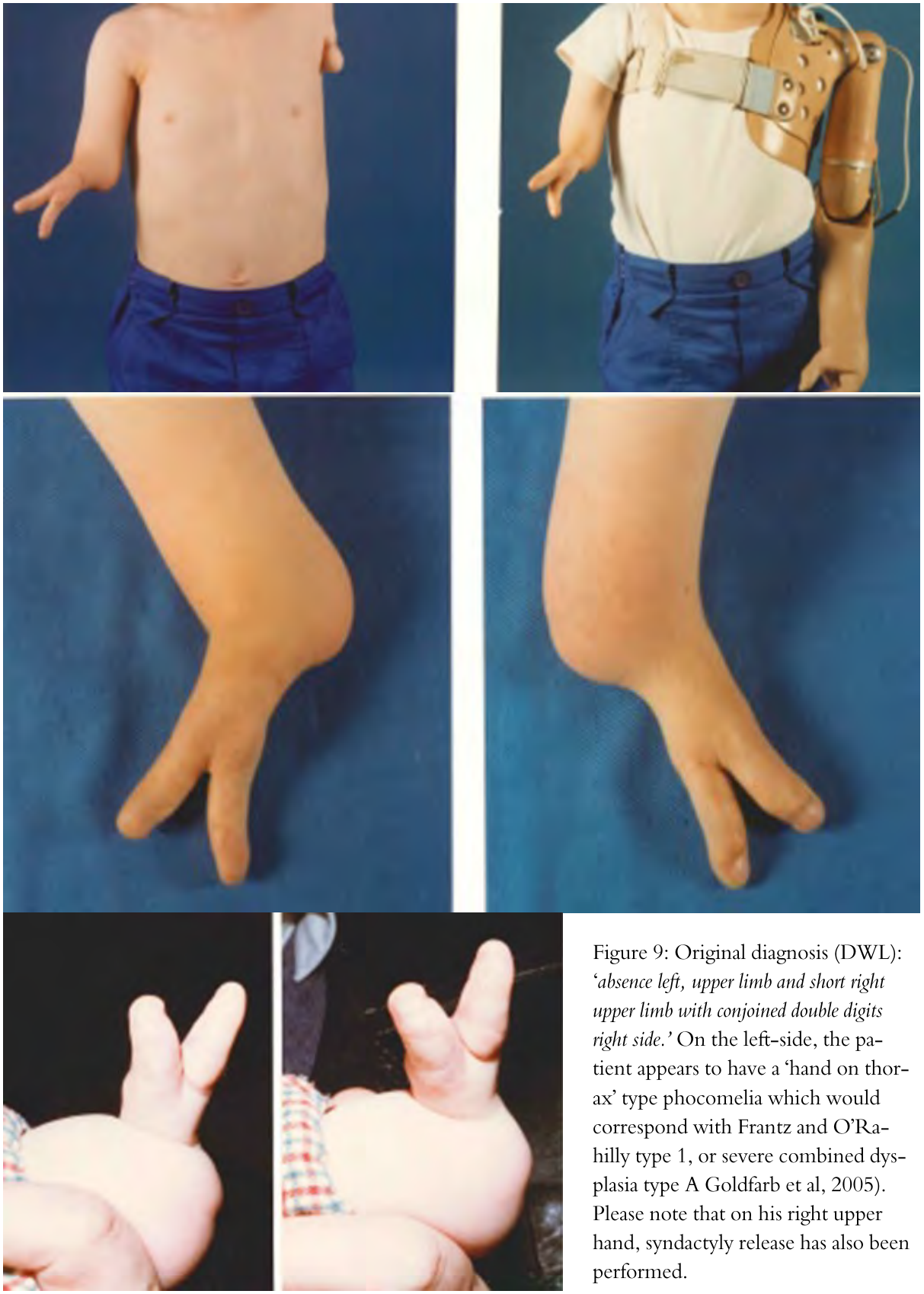


Figure 9: Original diagnosis (DWL): '*absence left, upper limb and short right upper limb with conjoined double digits right side.*' On the left-side, the patient appears to have a 'hand on thorax' type phocomelia which would correspond with Frantz and O'Rahilly type 1, or severe combined dysplasia type A Goldfarb et al, 2005). Please note that on his right upper hand, syndactyly release has also been performed.

CASE 8: *Phocomelia*

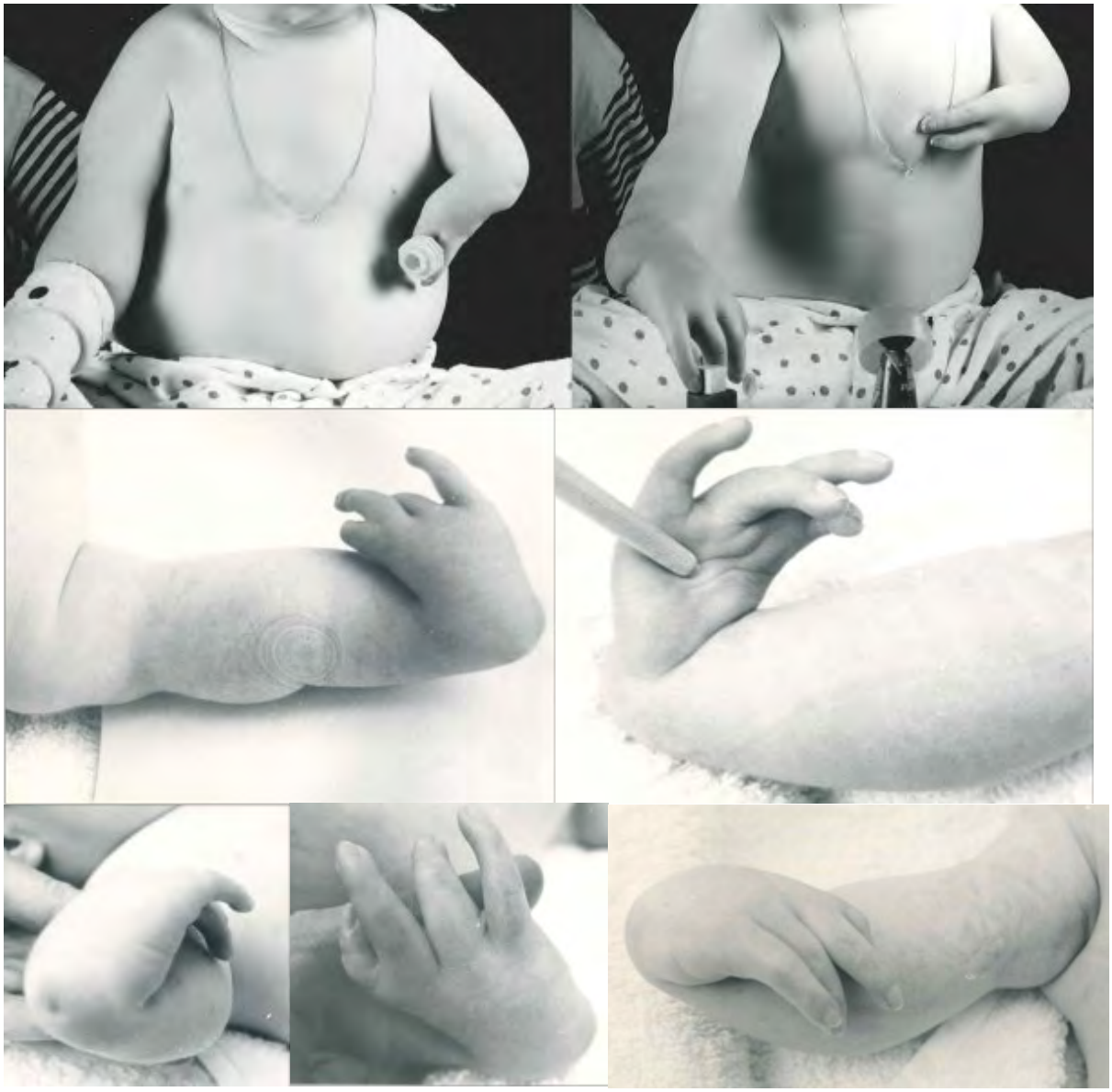


Figure 10: Original diagnosis (DWL): *'right radial club hand, and absence of humerus and radius with a 2-digit hand.'* It would appear that the patient has a shortened humerus with a very short forearm. This malformation would be classified as a severe proximal radial or ulnar dysplasia type condition Goldfarb et al, (2005) and not classifiable according to Frantz and O'Rahilly's classification.

CASE 9: *Phocomelia*

Figure 11: Original diagnosis (DWL): *'congenital absence of both upper limbs.'* On closer examination, the patient appears to have a severely hypoplastic humerus and forearm bones on each side. The hand is clearly present but severely hypoplastic bilaterally. These are single digit hands. This malformation can be classified as severe proximal radial or ulnar dysplasia (Goldfarb et al, 2005) and non-classifiable according to Frantz and O'Rahilly's classification.

CASE 10: *Phocomelia*

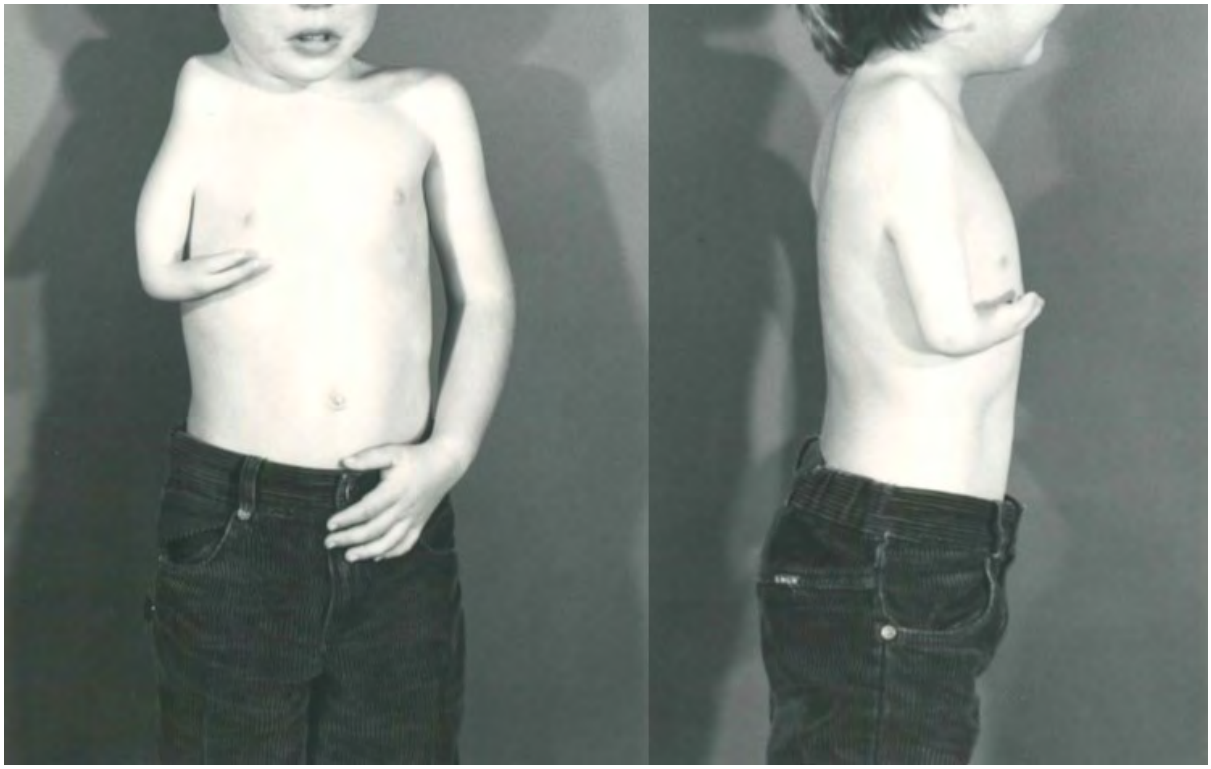


Figure 12: Original diagnosis (DWL): *'normal left upper limb. On the right, this patient has an abnormal shoulder girdle, without a forearm, with a humerus that is of near normal length and a hand directly attached to it*.' This would be considered a severe combined deficiency type A, or a Frantz and O'Rahilly type 2.

CASE 11: *Phocomelia*

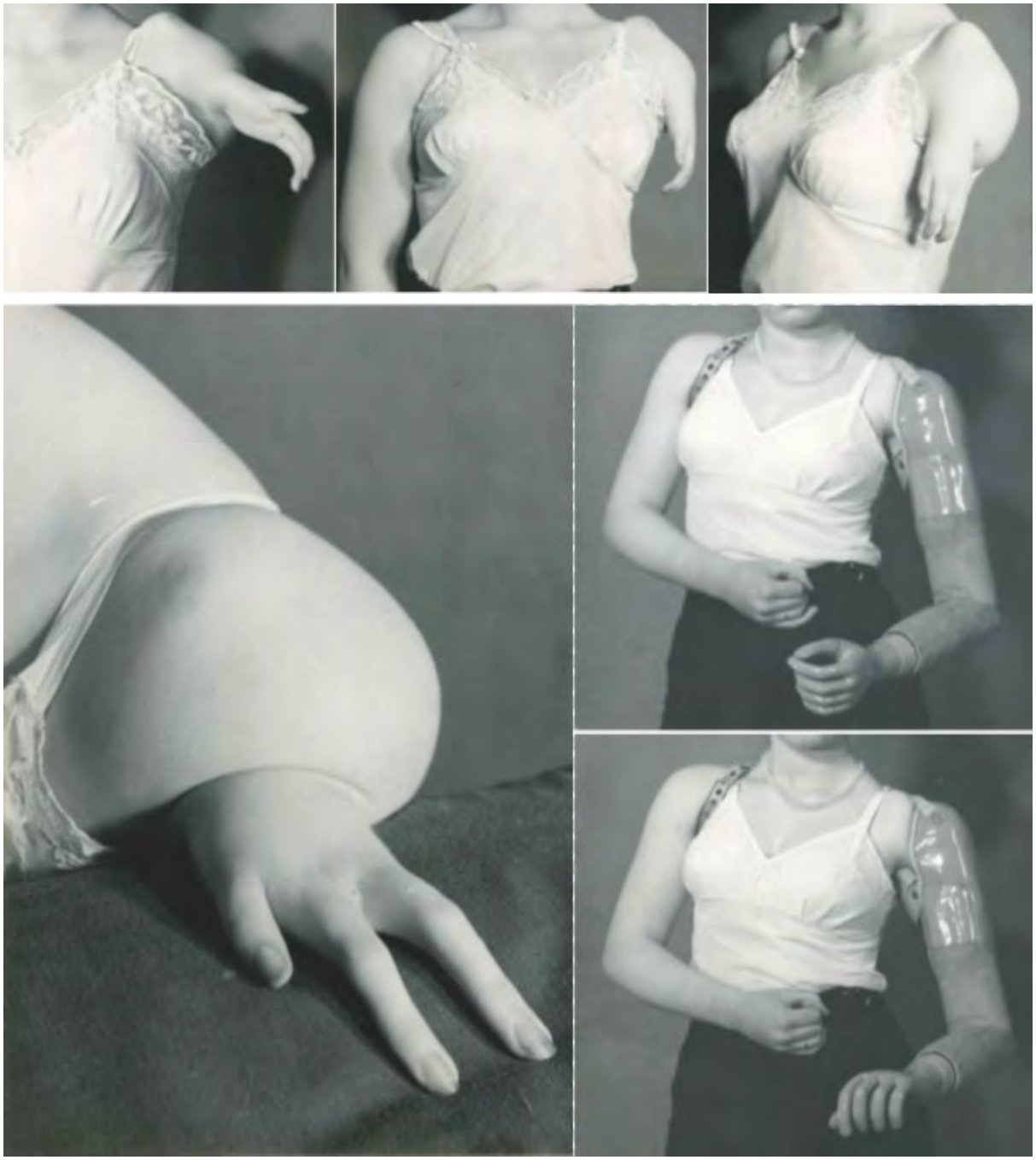


Figure 13: Original diagnosis (DWL): '*phocomelia*'. This patient has a 2-digit hand and a thumb that is directly attached to the thorax. This would correspond with a severe combined deficiency Type B (Goldfarb et al, 2005) and a Frantz and O'Rahilly Type 1.

CASE 12: *Phocomelia*

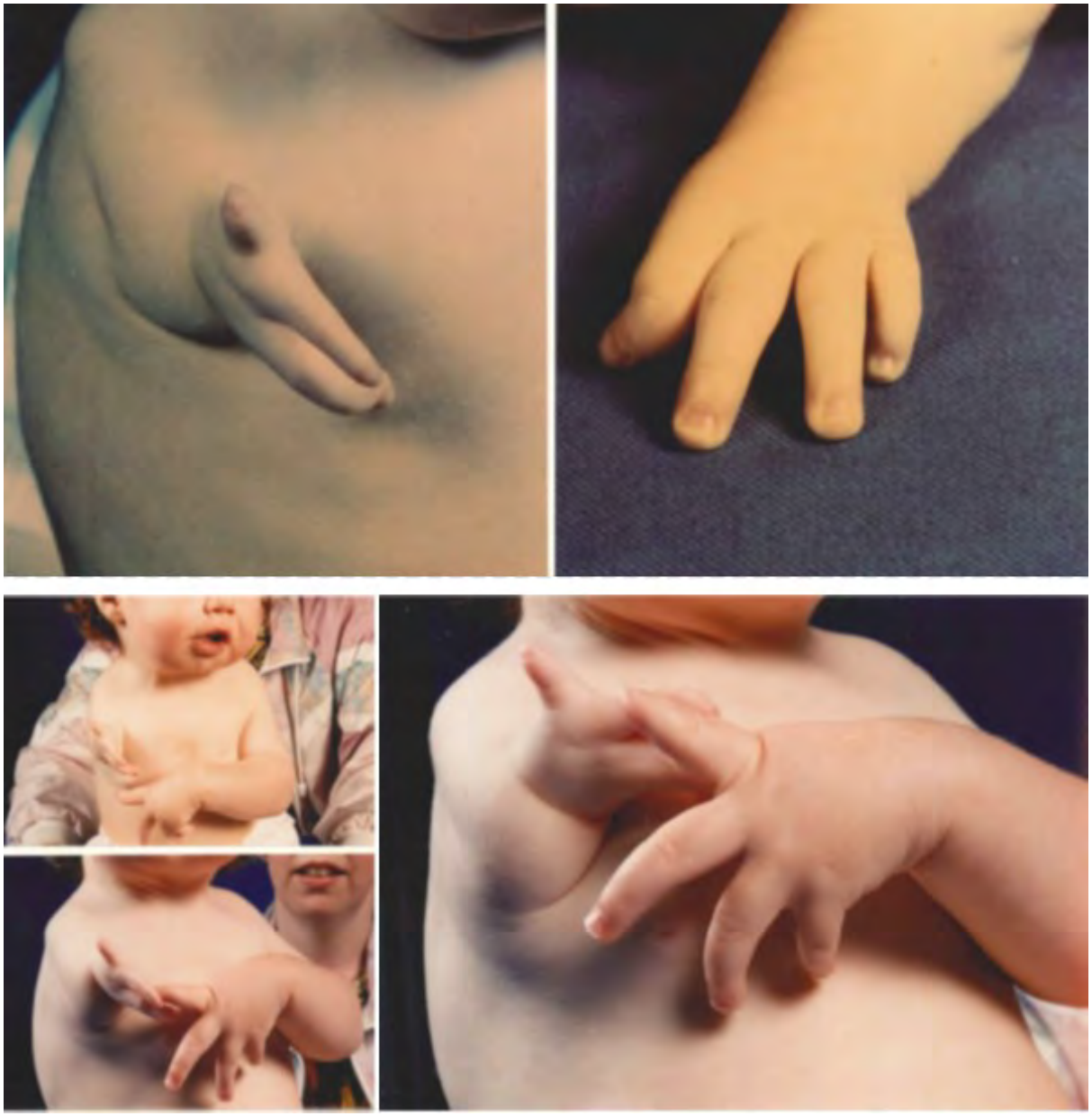


Figure 14: Original diagnosis (DWL): *'This young lady appears to have a near-normal left upper limb with a 4-digit hand.'* On the right-side, she has a hand on thorax condition which would correspond to a severe combined deficiency Type B (Goldfarb et al, 2005) and a Frantz and O'Rahilly Type 1.

The Evolution of Prostheses in Practice

IN HIS PRACTICE, there was a trend towards early fitting of prosthesis in many congenital hand conditions but especially in those affecting the entire limb, i.e., with a shortened upper arm or forearm. (Lamb et al., 1971). In this paper, they reviewed 67 patients with unilateral terminal absence of upper limb development. It seemed standard practice to fit a prosthesis initially on a child between the ages of 4 to 6 months. Prior to 1964, the earliest prosthetic fitting was made at the age of 4 years and in some cases a prosthesis had not been provided until the child reached the age of adolescence or later. Children were grouped according to the age at which they had their prosthesis fitted. In general, children who had prosthesis fitted earlier seemed to have developed active prehension with natural bimanual function under the age of 2 years, whereas children whose prosthesis was fitted much later (Group 4) until after the age of 5, presented with more problems especially related to prosthetic intolerance.

The practice of prosthetic fitting appears to have changed considerably over the years. Children may be considered for prosthetic fitting following referral to a congenital hand clinic or directly to a prosthetic service, but in general, routine prosthetic fitting is not practised until they are much older. Routine care is generally under the care of the prosthetist and surgeon. A surgeon would seldom see a child just to check fitting, or to make recommendations about the type of prosthesis. Perhaps there is much we can learn from these earlier experiences in terms of follow-up and the close input of surgeons surrounding prosthetic advice and ongoing care.

I-A-1. Malformation of the Entire Upper Limb: ***Radioulnar Axis***

THE RADIOULNAR AXIS is also known as the anterior posterior axis of development. This is controlled by the zone polarising activity or ZPA situated on the posterior limb bud, and the responsible protein is the sonic hedgehog (SHH), secreted in an ulnarising gradient of diffusion. Together with the proximal-distal axis, the radial-ulnar axis controls digit growth and also digit patterning, as well as the formation and identity of the forearm structures. It is generally thought that the ulna forms first, followed by the radius. Pathologies in this axis, usually via aberrant long range control of SHH activity especially in the earlier stages of gestation, (Lam et al., 2019) would give rise to the condition of radial longitudinal deficiency, otherwise known as the radial club hand. Radial longitudinal deficiency can occur in isolation but also in association with other syndromes including VACTERL, Holt-Oram, Fanconi anaemia and thrombocytopenia with absence of the radius (TAR).

Ulnar longitudinal deficiency, when the ulnar is found to be absent or short, is thought to be the result of a deficiency of the SHH protein. For this reason, ulnar longitudinal deficiency is rarely associated with other visceral malformations, unlike radial longitudinal deficiency, as children who survived to term probably had minimal SHH deficiencies to affect ulnar-sided (or fibula-sided) formation only.

Partly as a result of the thalidomide tragedy in the 1960s, Douglas Lamb and team saw a large number of children with radial longitudinal deficiency. He described the operation of 'centralization', although this term has since been used loosely with any procedure that centralized the wrist over the ulna. In his original description (Lamb, 1977) Douglas described the creation of a carpal slot, the depth of which must equal the diameter of the ulna and that the ulna must fit snugly and easily into the slot. Perhaps to avoid any stress to the fragile physis, a K- wire is inserted in a proximal-to-distal direction through the shaft of the third or fourth metacarpal and out through the metacarpal head. It is then drilled proximally through the centre of the distal ulnar epiphysis and up the medullary cavity of the ulna. Tendon transfers were not routinely performed, but the most commonly described in his paper was the transfer of the flexor carpi radialis onto the dorsum of the wrist, and he also described the need for flexor tendon lengthening, predominantly the flexor digitorum superficialis after the wrist is straightened. Douglas frequently performed pollicization, if he considered its muscle and joint structures were sufficient to provide good function as a thumb. He would perform pollicizations bilaterally if the conditions were right, going against popular teaching at that time to only perform this in the dominant hand. In the longer term follow-up of these patients (Lamb et al, 1997) he found no cases of premature fusions, indicating that care taken to avoid undue stress on the ulnar physis had paid off. Recurrence was minimal, especially in those who had undergone spontaneous ulnocarpal fusion. The overall length was around 66% of the contralateral limb. The use of the pollicized digit, however, was more inconsistent, with some patients adopting an ulnar prehension despite having undergone centralization and pollicization.

CASE 13: *Radial Longitudinal Dysplasia*

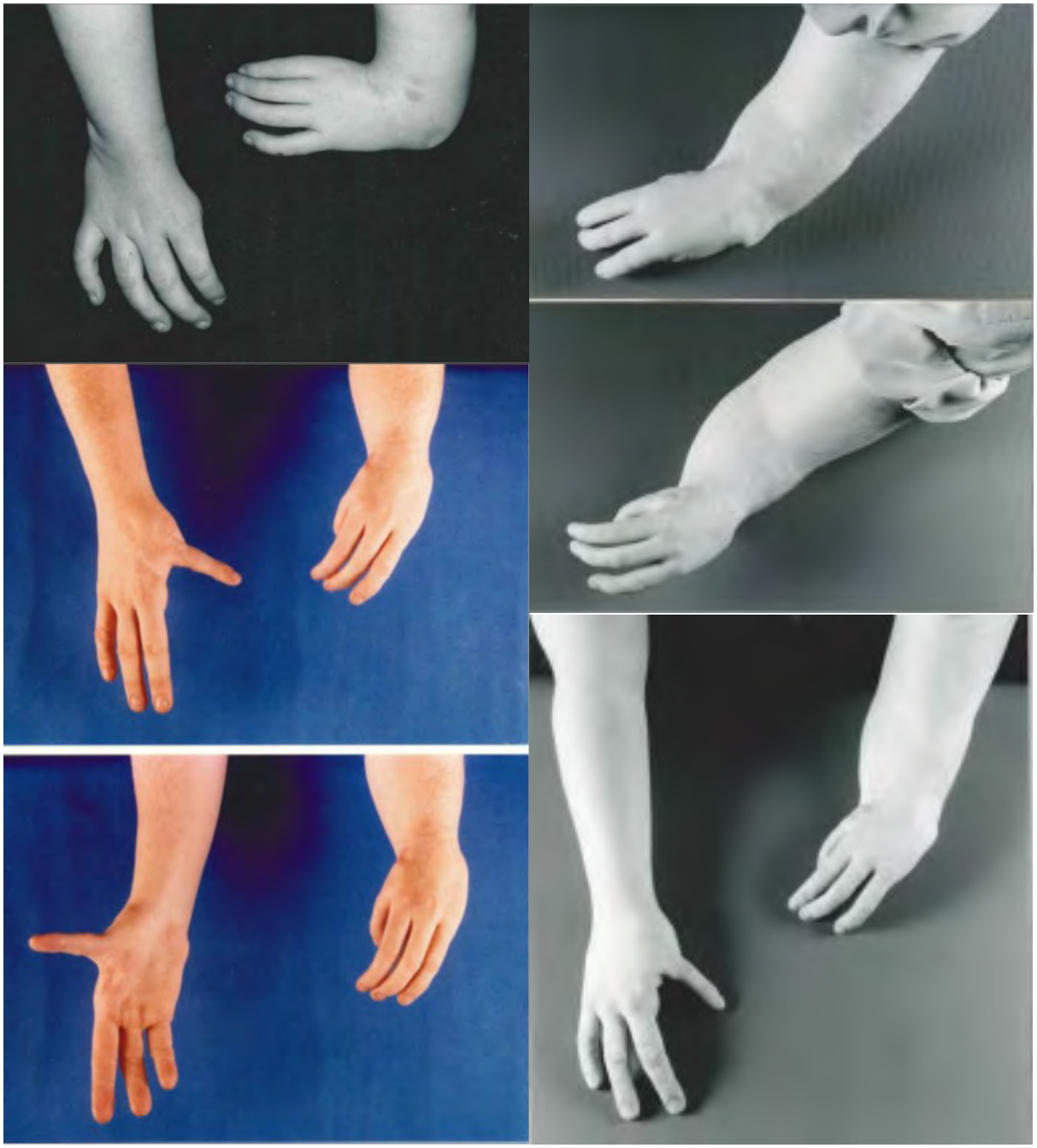


Figure 15: This patient has a left radial dysplasia and underwent the procedure of centralisation using a carpal slot. Post-operatively, the wrist is much straighter. However, she was still fitted with a prosthesis later on despite this improvement.

CASE 14: *Radial Longitudinal Dysplasia*

CLINICAL HISTORY

11.10.85

Seen at clinic by Mr Lamb

This child was brought upfrom theDurham area. He has an absence of the right radius with very marked deformity. This is very rigid and quite incorrectible. There is a stiff elbow and a very poor hand. There would certainly be very little to be gained functionally from any operation here. I do not think any corrective splintage would help here and correction would only be obtained which would not be justifiable with a stiff elbow and such a bad hand. I am more concerned about the torticollis which he exhibits. The head is held right over to the right. It is impossible to get full correction.

See 3 months.
Letter to Mr Checketts
DWL/AA

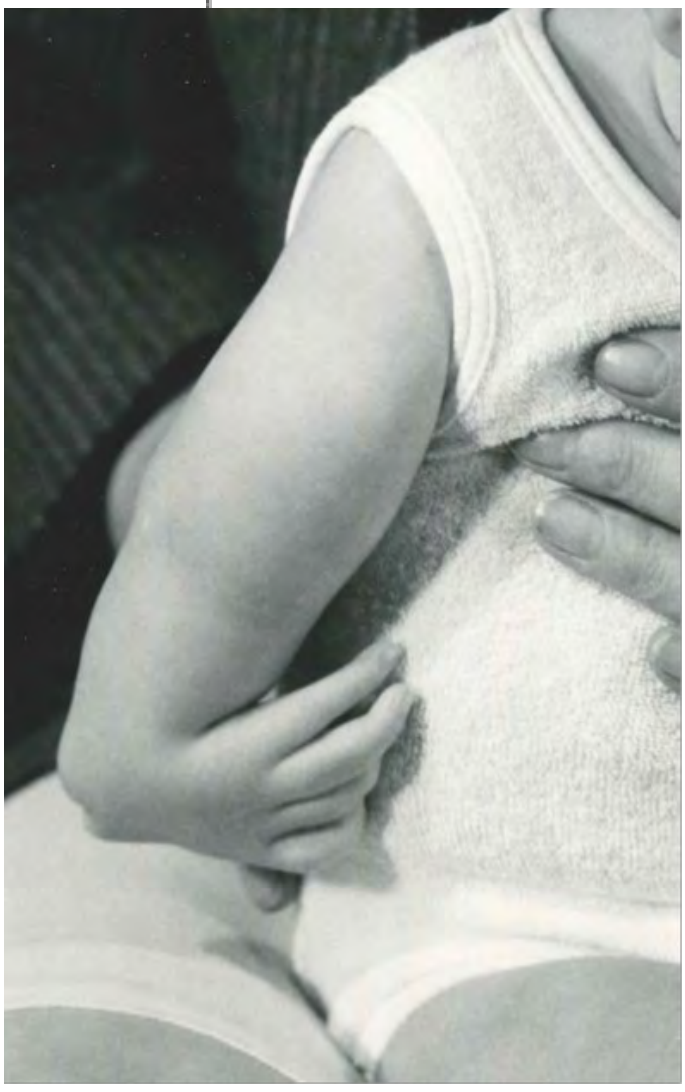


Figure 16: This patient has a right radial dysplasia with an absent thumb. He has a stiff elbow and therefore no centralisation was performed. Douglas was extremely strict about the contra-indications for centralisation, with the lack of elbow flexion being the main one. Any patients with a stiff elbow who underwent a centralisation procedure may result in the hand being unable to reach the mouth.

CASE 15: *Radial Longitudinal Dysplasia*

Seen at clinic by Mr Lamb

DIAGNOSIS : Bilat. radial club hands

HISTORY :

This child was referred by Mr Swanson, Senior Lecturer, Dundee. She is now 6 months old.

She has bilat. radial club hand deformity. On the right side there is no radius and on the left side hypoplastic radius. Spints have been worn night and day. The deformity is easily correctable passively. I think the splints could be worn at night only now.

On both hands the radial two digits are stiff at the PIPJ more marked on the right side. There appears maybe to be FDP ob both indices. There is good elbow movement. No other congenital abnormalities have been noted.

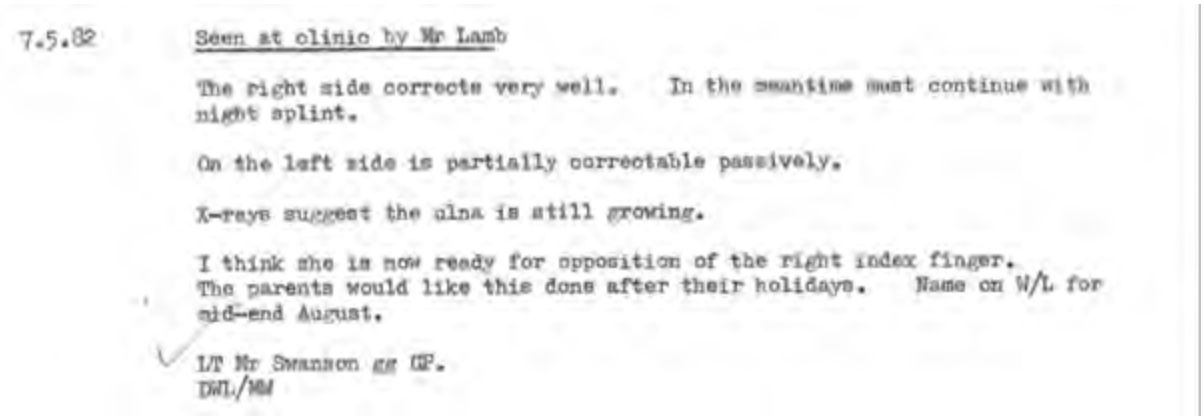
I think this girl would benefit from centralisation and probable pollicisation at least on one side. The left side would give the better function.

I have told the parents that I will write to Mr Swanson. We will wait to hear from him if he wishes me to deal with this.

L/T Mr Swanson cc GP.
DWL/MN

Figure 17: A child with bilateral radial dysplasia. On the right, he had a centralisation and pollicisation.

CASE 15 (continued): *Radial Longitudinal Dysplasia*



7.5.82 Seen at clinic by Mr Lamb

The right side corrects very well. In the meantime must continue with night splint.

On the left side is partially correctable passively.

X-rays suggest the ulna is still growing.

I think she is now ready for opposition of the right index finger. The parents would like this done after their holidays. Name on W/L for mid-end August.

LT Mr Swanson ~~gg~~ GP.
DWL/MM

Figure 18: Post-operative clinical notes after centralisation of the right wrist

25.11.82

Operation RIGHT INDEX POLLICISATION

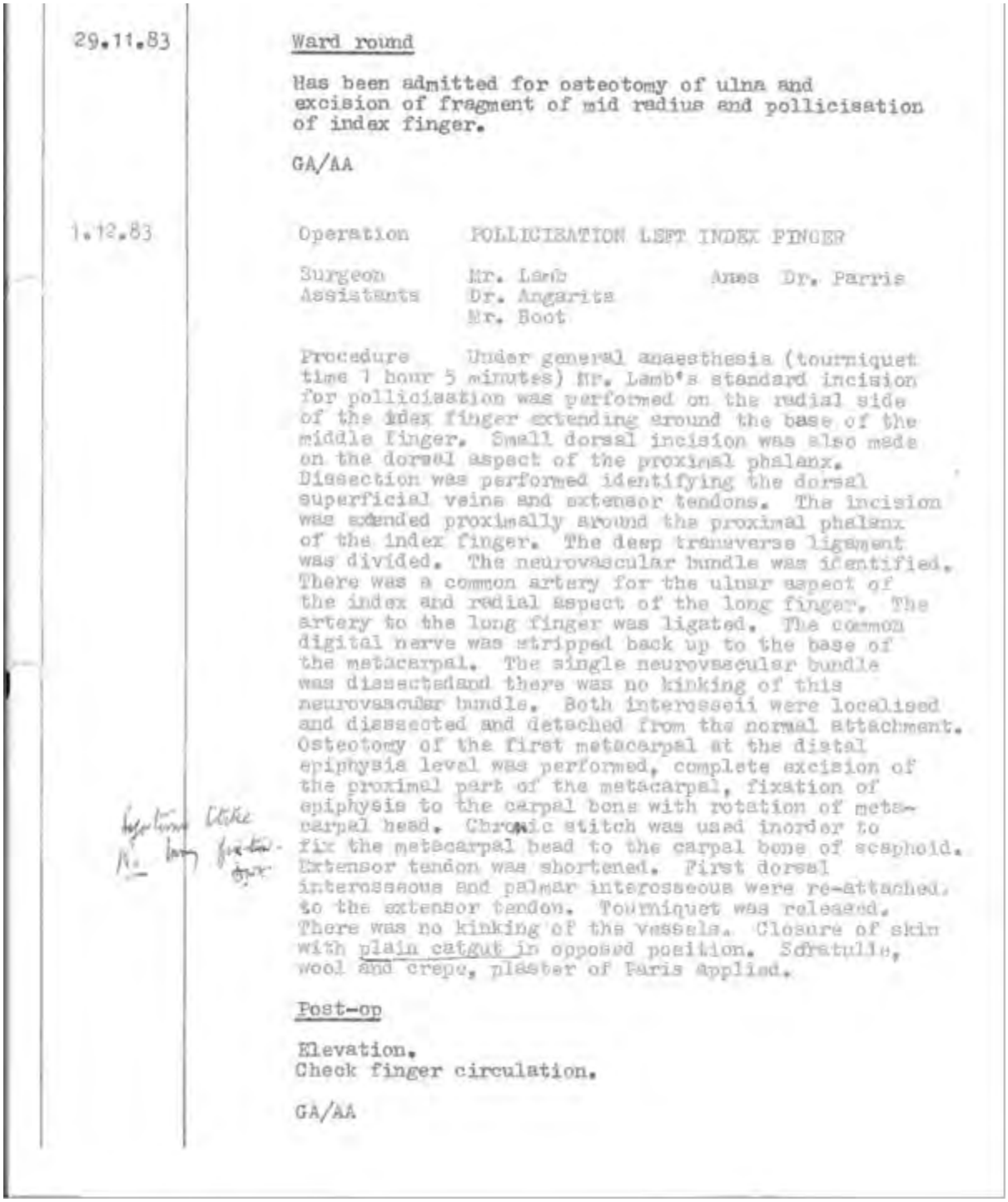
Surgeon Mr. Lamb Anaes Dr. Park
Assistant Mr. Hutchins GA T.T. 1½ hours

Procedure Skin flaps were developed at the base of the index finger using a racket incision, prolonged on to the dorso-radial aspect of the hand through a sinuous incision. There was a further prolongation of the incision on to the dorsum of the proximal phalanx. Skin flaps were developed and the anterior neurovascular bundles identified. The cleft between the second and third metacarpals was then developed and the digital artery identified. The radial digital vessels to the middle finger were ligated and divided. The dorsal venous pedicle was preserved. Subperiosteal dissection of the shaft of the metacarpal was then performed and the proximal portion of the metacarpal was excised through the epiphyseal plate. The intrinsic muscles were detached distally and retracted proximally. Three 30 chromic catgut sutures were then placed through the bed of the second metacarpal and through the head of the metacarpal so as to rotate it through 150 degrees and flex it palmarly. The new carpal bone was thus sunk into its new bed in the hand. The intrinsic muscles were then re-attached - the first dorsal interosseus to the periosteum of the new metacarpal, just proximal to the joint and the first volar interosseus to the extensor mechanism. The tourniquet was released with good return of circulation to the thumb. Skin flaps were closed with interrupted plain catgut sutures and a well padded dressing was applied with an overlying plaster slab to keep the new thumb in opposition.

PH/AA

Figure 19: Surgery notes for right index pollicisation

CASE 15 (continued): *Radial Longitudinal Dysplasia*



29.11.83

Ward round

Has been admitted for osteotomy of ulna and excision of fragment of mid radius and pollicisation of index finger.

GA/AA

1.12.83

Operation POLLICISATION LEFT INDEX FINGER

Surgeon Mr. Lamb Anes Dr. Parris
Assistants Dr. Angarita
Mr. Boot

Procedure Under general anaesthesia (tourniquet time 1 hour 5 minutes) Mr. Lamb's standard incision for pollicisation was performed on the radial side of the index finger extending around the base of the middle finger. Small dorsal incision was also made on the dorsal aspect of the proximal phalanx. Dissection was performed identifying the dorsal superficial veins and extensor tendons. The incision was extended proximally around the proximal phalanx of the index finger. The deep transverse ligament was divided. The neurovascular bundle was identified. There was a common artery for the ulnar aspect of the index and radial aspect of the long finger. The artery to the long finger was ligated. The common digital nerve was stripped back up to the base of the metacarpal. The single neurovascular bundle was dissectedand there was no kinking of this neurovascular bundle. Both interossei were localised and dissected and detached from the normal attachment. Osteotomy of the first metacarpal at the distal epiphysis level was performed, complete excision of the proximal part of the metacarpal, fixation of epiphysis to the carpal bons with rotation of metacarpal head. Chromic stitch was used inorder to fix the metacarpal head to the carpal bone of scaphoid. Extensor tendon was shortened. First dorsal interosseous and palmar interosseous were re-attached to the extensor tendon. Tourniquet was released. There was no kinking of the vessels. Closure of skin with plain catgut in opposed position. Sofratulle, wool and crepe, plaster of Paris applied.

Post-op

Elevation.
Check finger circulation.

GA/AA

Figure 20: Surgery notes for left index pollicisation

CASE 16: *Radial Longitudinal Dysplasia*

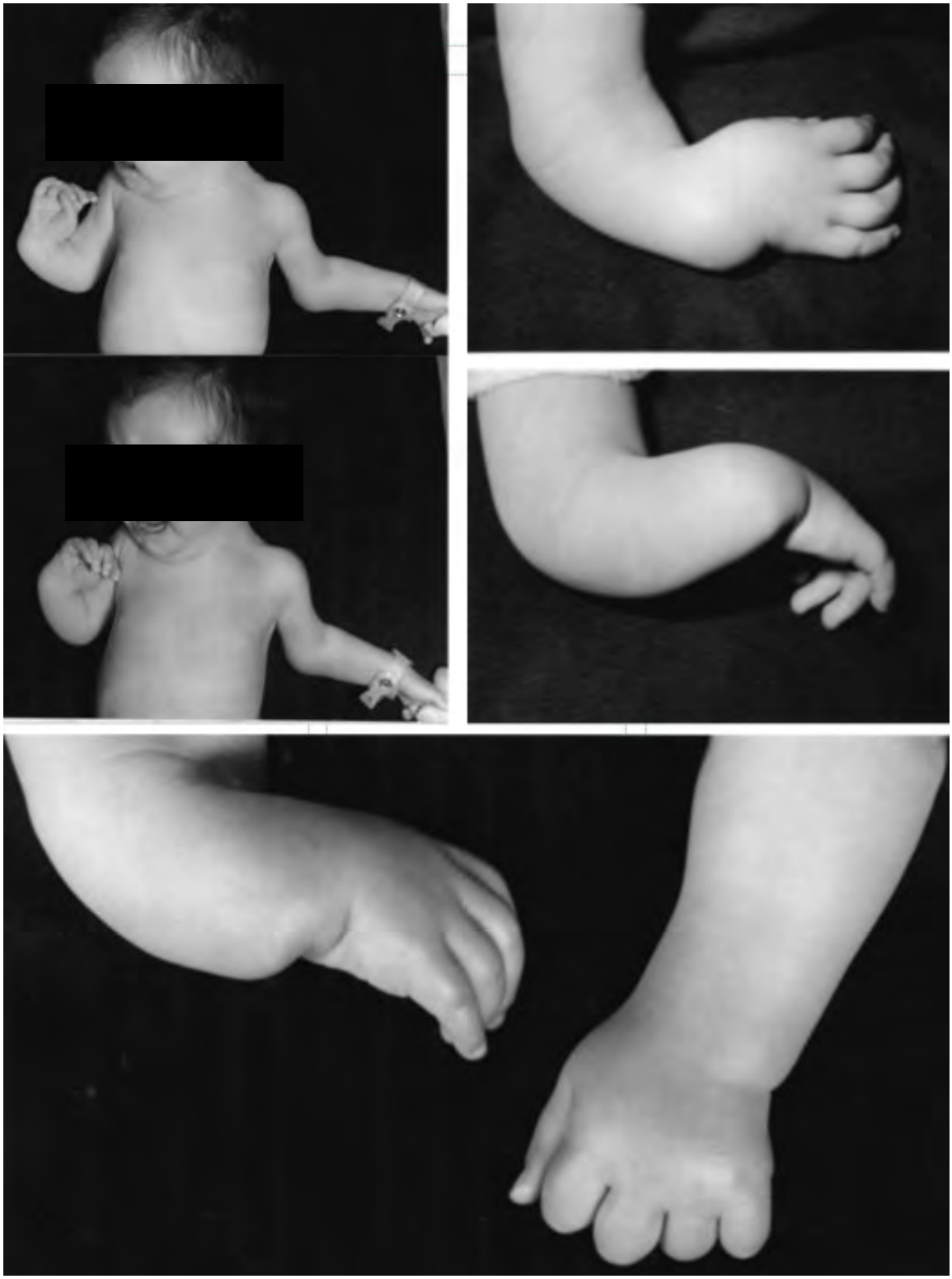
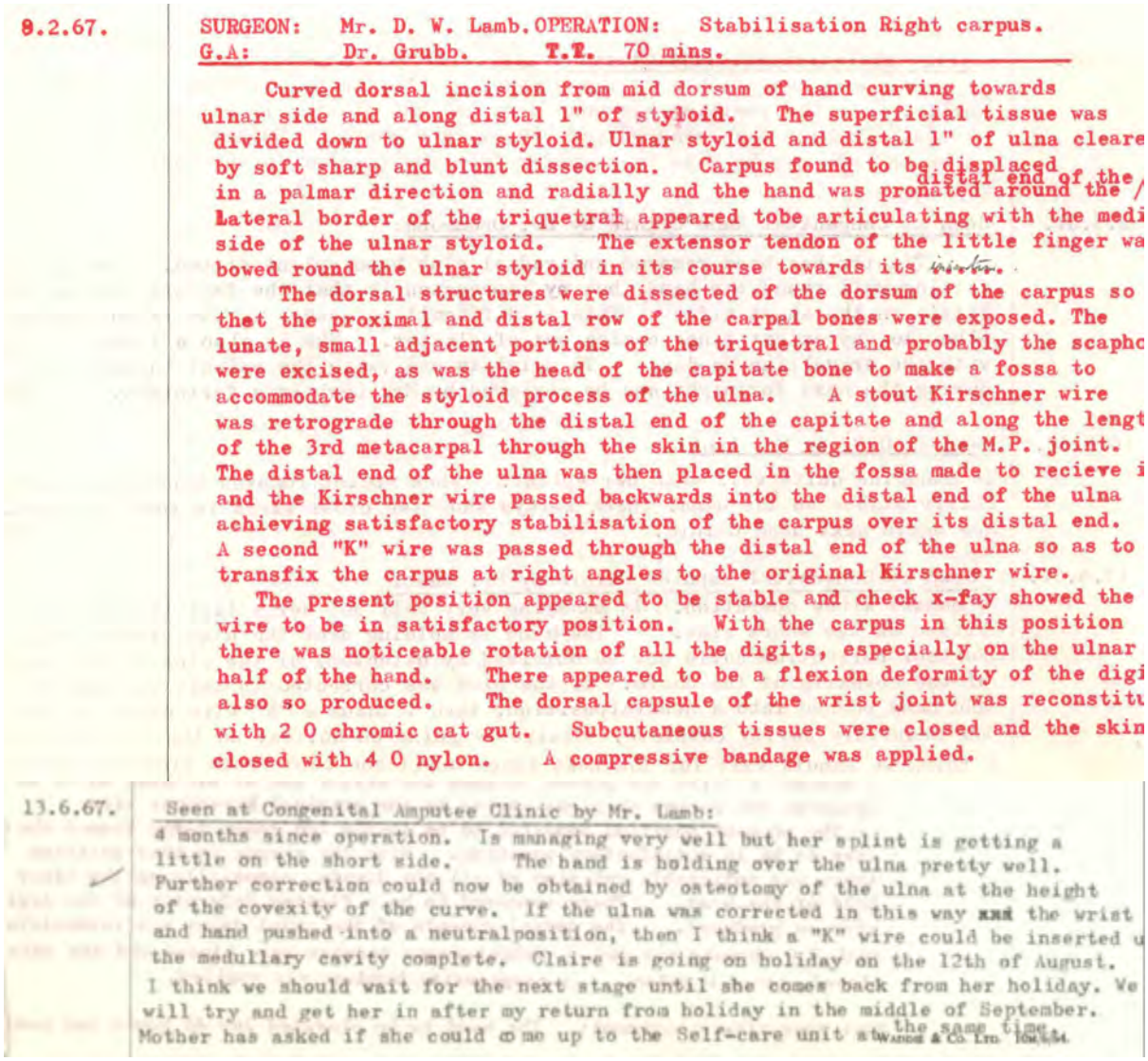


Figure 21: A child with right radial dysplasia and hypoplastic thumb. She underwent a right centralization with a carpal slot.

CASE 16 (continued): *Radial Longitudinal Dysplasia*



9.2.67.

SURGEON: Mr. D. W. Lamb. OPERATION: Stabilisation Right carpus.
G.A: Dr. Grubb. T.T. 70 mins.

Curved dorsal incision from mid dorsum of hand curving towards ulnar side and along distal 1" of styloid. The superficial tissue was divided down to ulnar styloid. Ulnar styloid and distal 1" of ulna cleare by soft sharp and blunt dissection. Carpus found to be displaced in a palmar direction and radially and the hand was pronated around the distal end of the / Lateral border of the triquetral appeared tobe articulating with the medi side of the ulnar styloid. The extensor tendon of the little finger wa bowed round the ulnar styloid in its course towards its ~~ulnar~~.

The dorsal structures were dissected of the dorsum of the carpus so that the proximal and distal row of the carpal bones were exposed. The lunate, small adjacent portions of the triquetral and probably the scapho were excised, as was the head of the capitate bone to make a fossa to accommodate the styloid process of the ulna. A stout Kirschner wire was retrograde through the distal end of the capitate and along the lengt of the 3rd metacarpal through the skin in the region of the M.P. joint. The distal end of the ulna was then placed in the fossa made to recieve i and the Kirschner wire passed backwards into the distal end of the ulna achieving satisfactory stabilisation of the carpus over its distal end. A second "K" wire was passed through the distal end of the ulna so as to transfix the carpus at right angles to the original Kirschner wire.

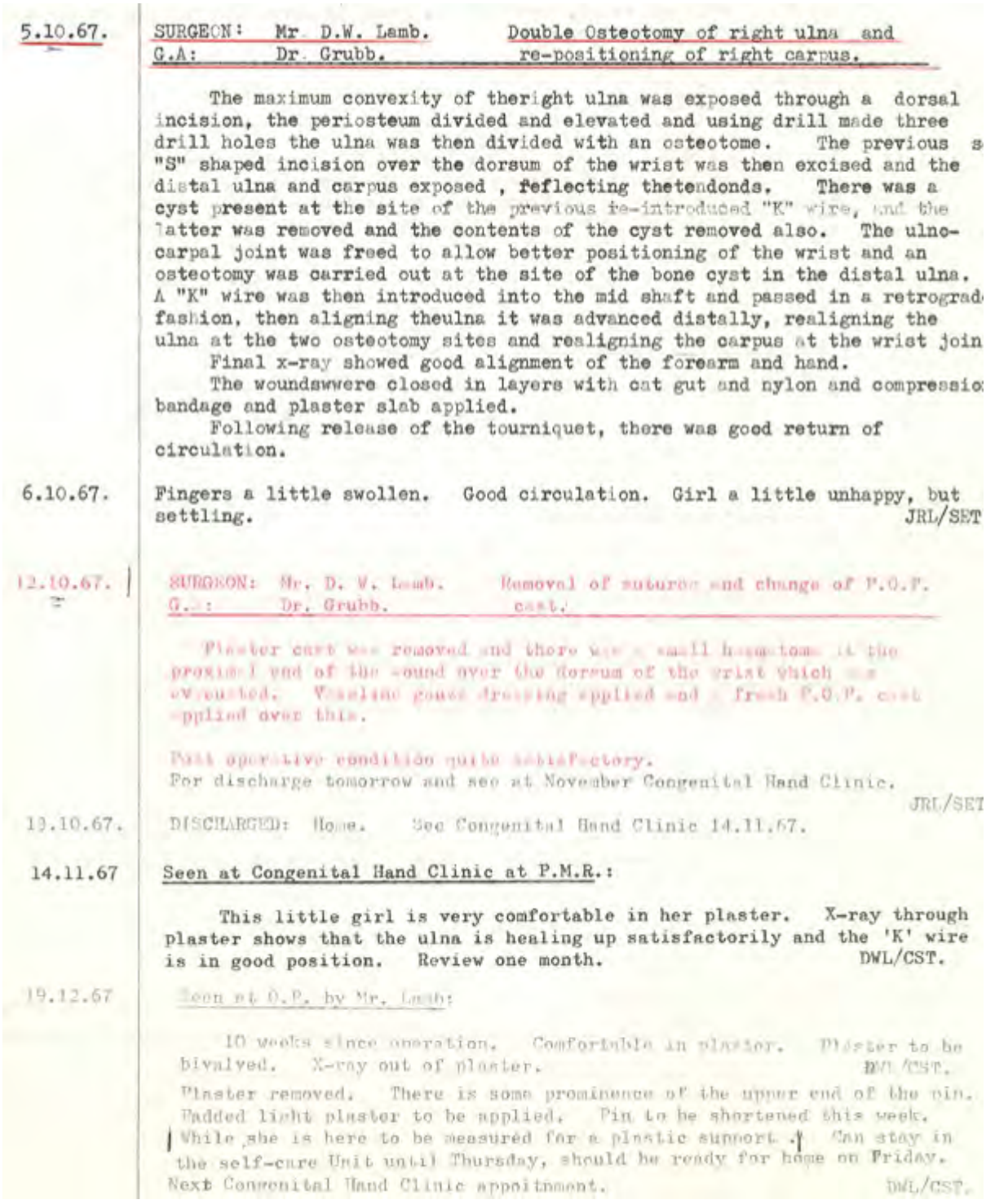
The present position appeared to be stable and check x-fay showed the wire to be in satisfactory position. With the carpus in this position there was noticeable rotation of all the digits, especially on the ulnar half of the hand. There appeared to be a flexion deformity of the digi also so produced. The dorsal capsule of the wrist joint was reconstitu with 2 0 chromic cat gut. Subcutaneous tissues were closed and the skin closed with 4 0 nylon. A compressive bandage was applied.

13.6.67.

Seen at Congenital Amputee Clinic by Mr. Lamb:
4 months since operation. Is managing very well but her splint is getting a little on the short side. The hand is holding over the ulna pretty well. Further correction could now be obtained by osteotomy of the ulna at the height of the covexity of the curve. If the ulna was corrected in this way ~~and~~ the wrist and hand pushed into a neutralposition, then I think a "K" wire could be inserted u the medullary cavity complete. Claire is going on holiday on the 12th of August. I think we should wait for the next stage until she comes back from her holiday. We will try and get her in after my return from holiday in the middle of September. Mother has asked if she could come up to the Self-care unit at the same time.

Figure 22: Surgery notes for right centralisation. Post-operatively, she was seen at 4 months and it was felt that further osteotomies could be beneficial to straighten the forearm.

CASE 16 (continued): *Radial Longitudinal Dysplasia*



5.10.67.

SURGEON: Mr. D.W. Lamb. Double Osteotomy of right ulna and
G.A: Dr. Grubb. re-positioning of right carpus.

The maximum convexity of theright ulna was exposed through a dorsal incision, the periosteum divided and elevated and using drill made three drill holes the ulna was then divided with an osteotome. The previous s "S" shaped incision over the dorsum of the wrist was then excised and the distal ulna and carpus exposed , reflecting thetendonds. There was a cyst present at the site of the previous re-introduced "K" wire, and the latter was removed and the contents of the cyst removed also. The ulno-carpal joint was freed to allow better positioning of the wrist and an osteotomy was carried out at the site of the bone cyst in the distal ulna. A "K" wire was then introduced into the mid shaft and passed in a retrograd fashion, then aligning theulna it was advanced distally, realigning the ulna at the two osteotomy sites and realigning the carpus at the wrist join
Final x-ray showed good alignment of the forearm and hand.
The woundswwere closed in layers with cat gut and nylon and compressio bandage and plaster slab applied.
Following release of the tourniquet, there was good return of circulation.

6.10.67. Fingers a little swollen. Good circulation. Girl a little unhappy, but settling. JRL/SET

12.10.67. SURGEON: Mr. D. W. Lamb. Removal of sutures and change of P.O.P.
G.A.: Dr. Grubb. cast.

Plaster cast was removed and there was a small haematoma at the proximal end of the wound over the dorsum of the wrist which was evacuated. Vaseline gauze dressing applied and a fresh P.O.P. cast applied over this.

Post operative condition quite satisfactory.
For discharge tomorrow and see at November Congenital Hand Clinic.
JRL/SET

13.10.67. DISCHARGED: Home. See Congenital Hand Clinic 14.11.67.

14.11.67 Seen at Congenital Hand Clinic at P.M.R.:

This little girl is very comfortable in her plaster. X-ray through plaster shows that the ulna is healing up satisfactorily and the 'K' wire is in good position. Review one month. DWL/CST.

19.12.67 Seen at O.P. by Mr. Lamb:

10 weeks since operation. Comfortable in plaster. Plaster to be bivalved. X-ray out of plaster. DWL/CST.
Plaster removed. There is some prominence of the upper end of the pin. Padded light plaster to be applied. Pin to be shortened this week. While she is here to be measured for a plastic support. Can stay in the self-care Unit until Thursday, should be ready for home on Friday. Next Congenital Hand Clinic appointment. DWL/CST.

Figure 23: Surgery notes for double osteotomy in the same patient and re-positioning of the carpus.

CASE 16 (continued): *Radial Longitudinal Dysplasia*

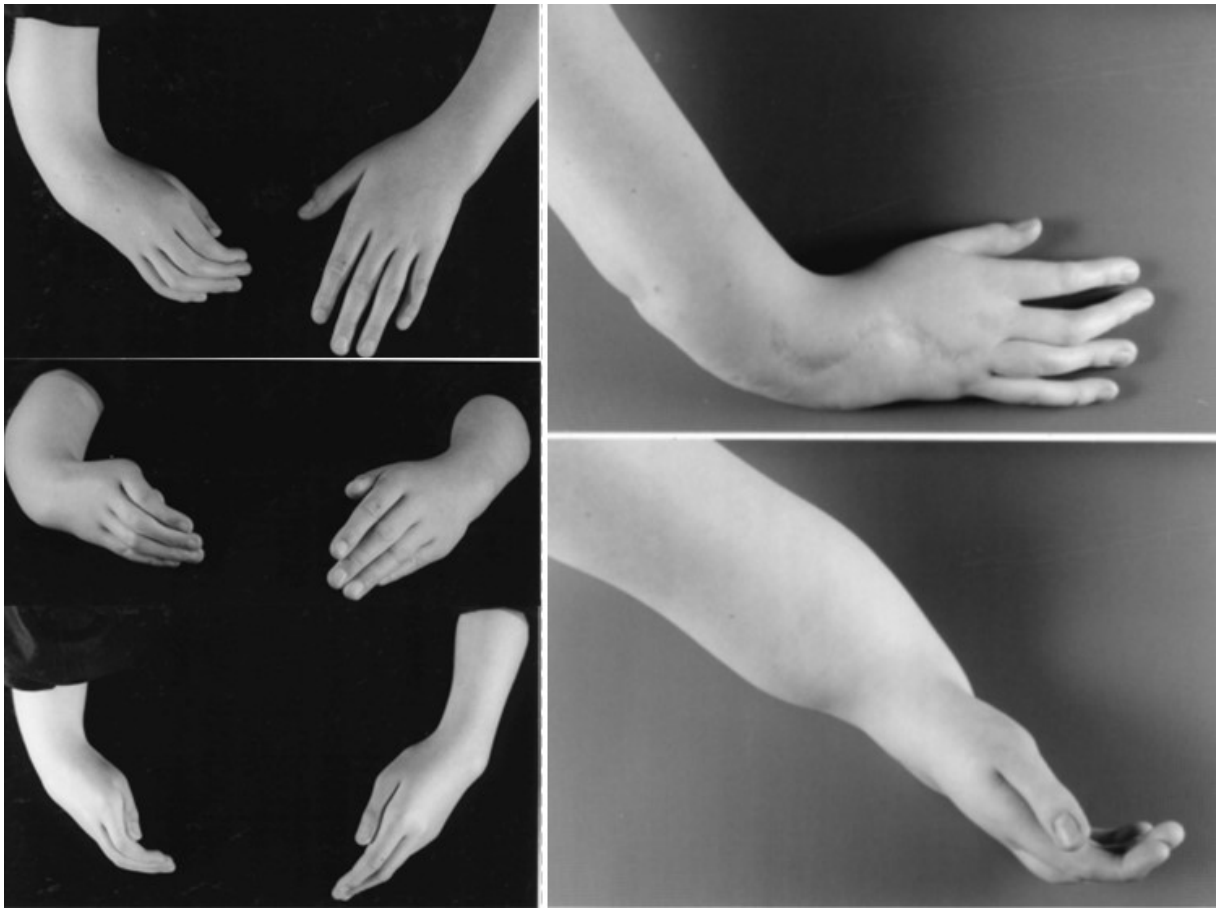


Figure 24: These photographs show the progression of recurrence in the same patient over time, following surgical correction. Initially, the wrist appears straight but over time the deviation recurred. The original subluxation appears to have been corrected effectively. .

CASE 17: *Radial Longitudinal Dysplasia*

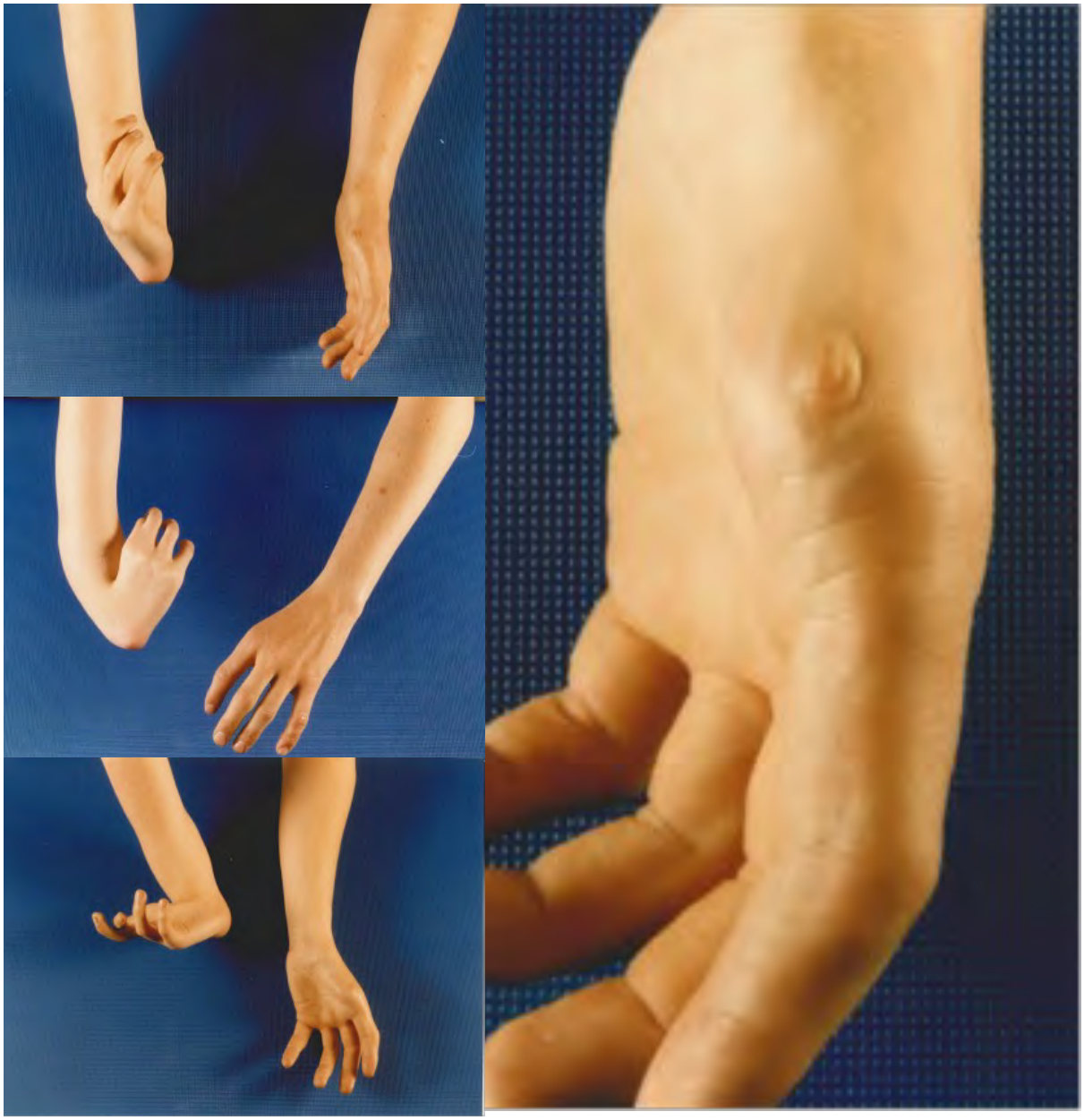


Figure 25: This patient has a severe right radial dysplasia and presented late at age 18. The deviation was more than 120° and with elbow stiffness. In this case, the decision was made to leave things as they were, and to wait until the right forearm stops growing and then consider a fusion to improve the appearance. (See Figure 26 for clinical notes on next page)

CASE 17 (continued): *Radial Longitudinal Dysplasia*

CLINICAL HISTORY

SEENA T CLINIC BY MR LAMB

DOAGNOSIS :

HISTORY :

See with mother and younger brother
Nice young man who is now becoming sensitive about the
appearance particularly of his right upper limb.
Cong absence of radious with marked club hand deformity which
is not apparently being controlled or corrected in any way.
It is now a very fixed contracture. He has stiff fingers
with very poor active and passive mvoement even on the ulna
digit. Uses the bent wrist as a hook to carry things.

Left upper limb normal apart from absence of the thumb.

Seen early on by George Lloyd-Roberts, advised not to have anythir
done. In retrospect I am sure this boy would ahve been
better with a pollicisation of the index on he left side
and centralisation on the right. However, I am sure it is
the right thing not to do anything now.

He would look better on the right side by having stabilisation
of the wrist but he still has a lower ulnar epiphysis which
is open and I think this should be left. Might consider
arthrodesis of the wrist subsequebntly.

I have explained this to boy and mother who I am sure accept this
and will write along these lines to Rolfe Birch.

Thank you for your letter about this lad and for asking me tos ee him.
I saw him today with his mother.

I am sure your decision against operation is the right one at this stage.
I think it would be most unwise to do any pollicisation of his left index
finger at this stage.

He is obviously getting concerned about the appearance of his short and
deformed right upper limb. I have stressed to mother that the cardinal
rule is to try and improve function and this is not possible at least to
do any harm to what function is available. Probably straightening his wrist
at this stage would not cause any deterioration in function but it certainly
would not improve it and the only gain would be in appearance. However
X-rays show that his lower ulnar epiphysis is still growing and I am sure
it would be wrong to do anything while the epiphysis is still open. Once
it is closed the boy must make up his own mind whether he would wish to
have the wrist corrected for appearances sake and it would certainly make
this very short forearm look that much longer.

If at that stage he did decide to ahve the wrist straightened I would not think
a centralisation would be appropriate and it would have to be a mroe formal
type of arthrodesis but getting correction is obviously going to be difficult
with this very tight contracture of skin and soft tissue.

With kind regards,

Yours sincerely

Figure 26: Clinical notes for the above patient with right radial dysplasia.

CASE 18: *Radial Longitudinal Dysplasia*

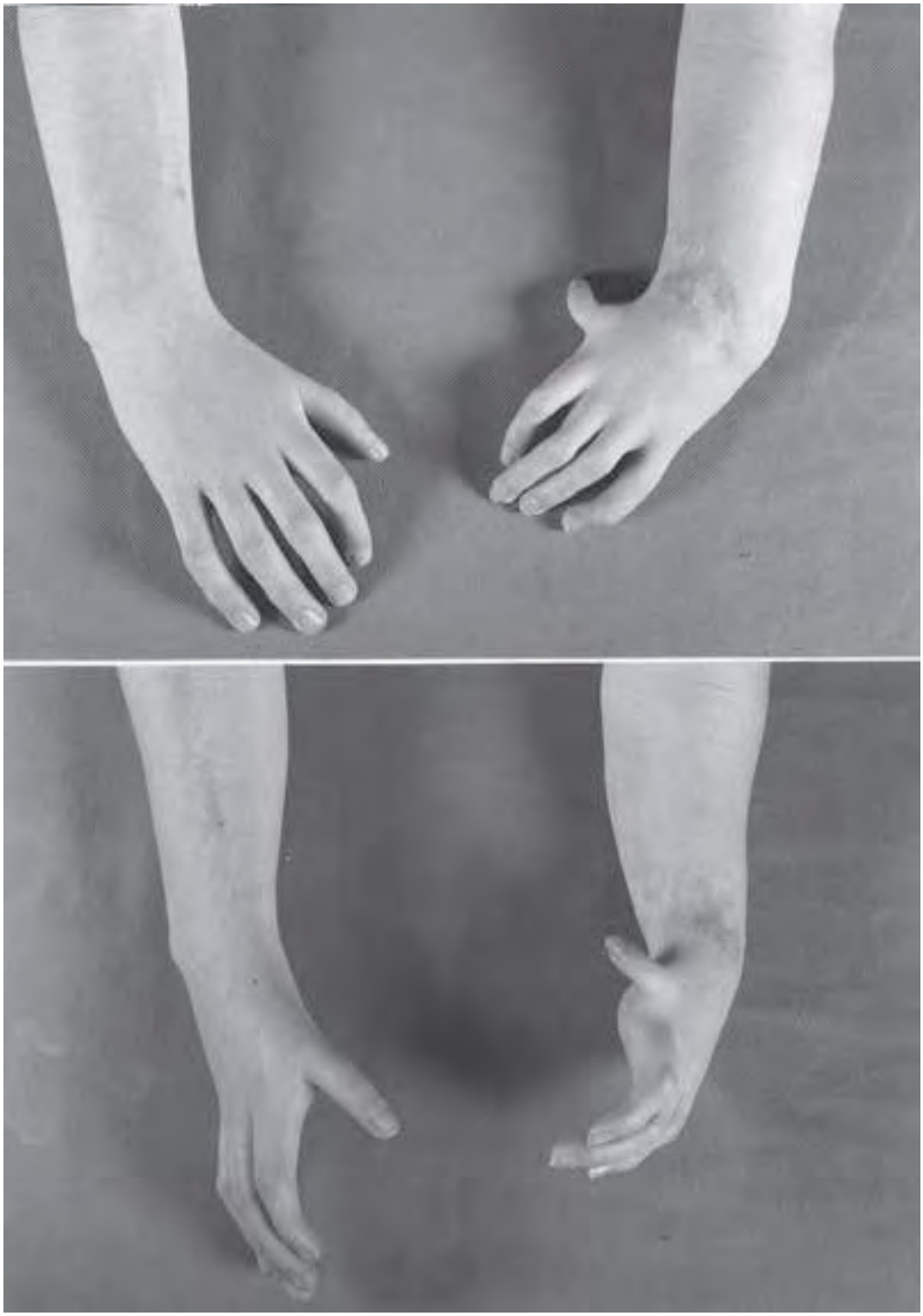
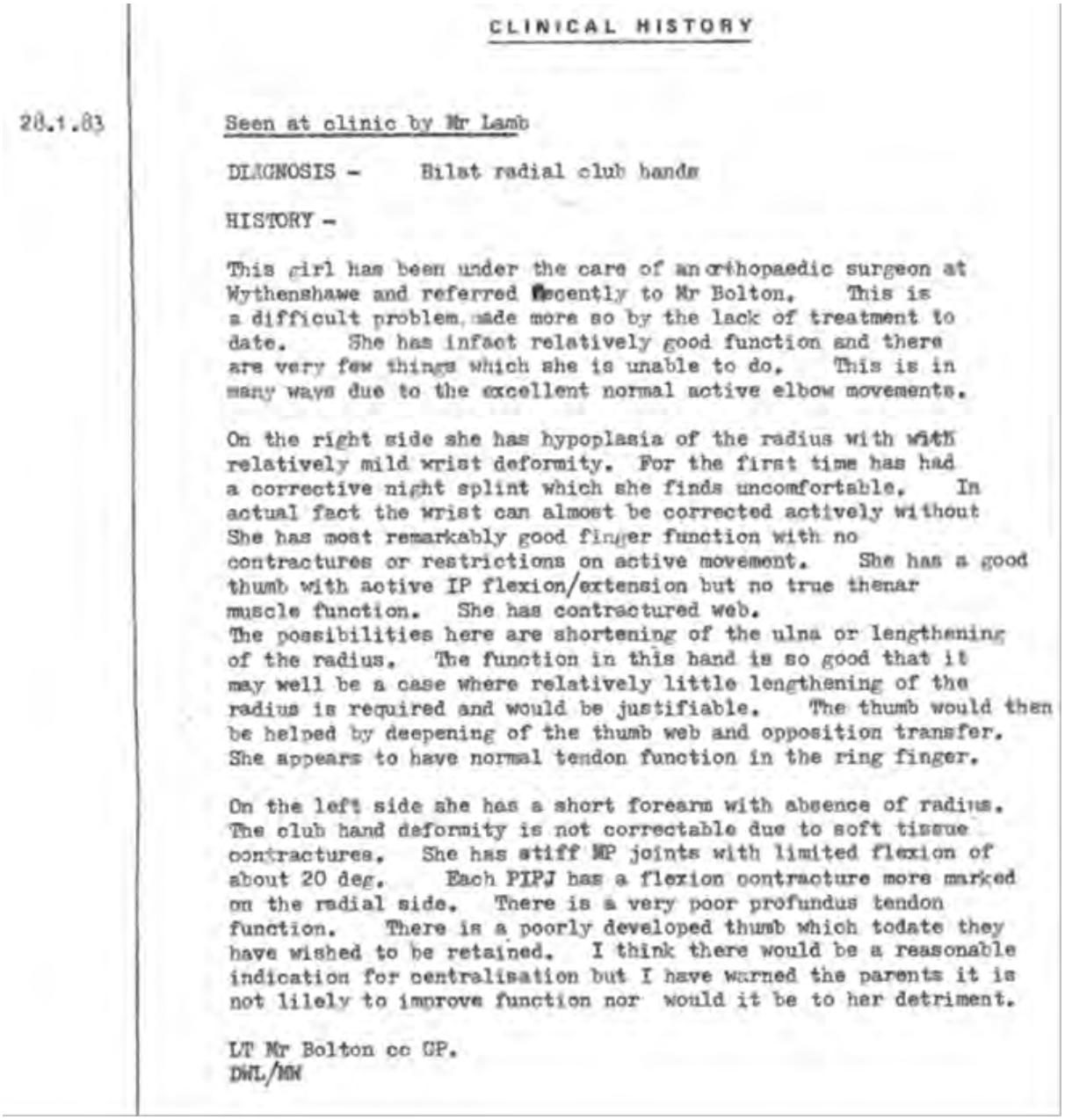


Figure 27: This patient has bilateral radial dysplasia: mild deviation on the right side with a hypoplastic radius and a sever left sided radial dysplasia with absent radius. She underwent lengthening of the right radius and centralization of the left wrist.

CASE 18 (continued): *Radial Longitudinal Dysplasia*



CLINICAL HISTORY

28.1.83

Seen at clinic by Mr Lamb

DIAGNOSIS - Bilat radial club hands

HISTORY -

This girl has been under the care of an orthopaedic surgeon at Wythenshawe and referred recently to Mr Bolton. This is a difficult problem, made more so by the lack of treatment to date. She has infact relatively good function and there are very few things which she is unable to do. This is in many ways due to the excellent normal active elbow movements.

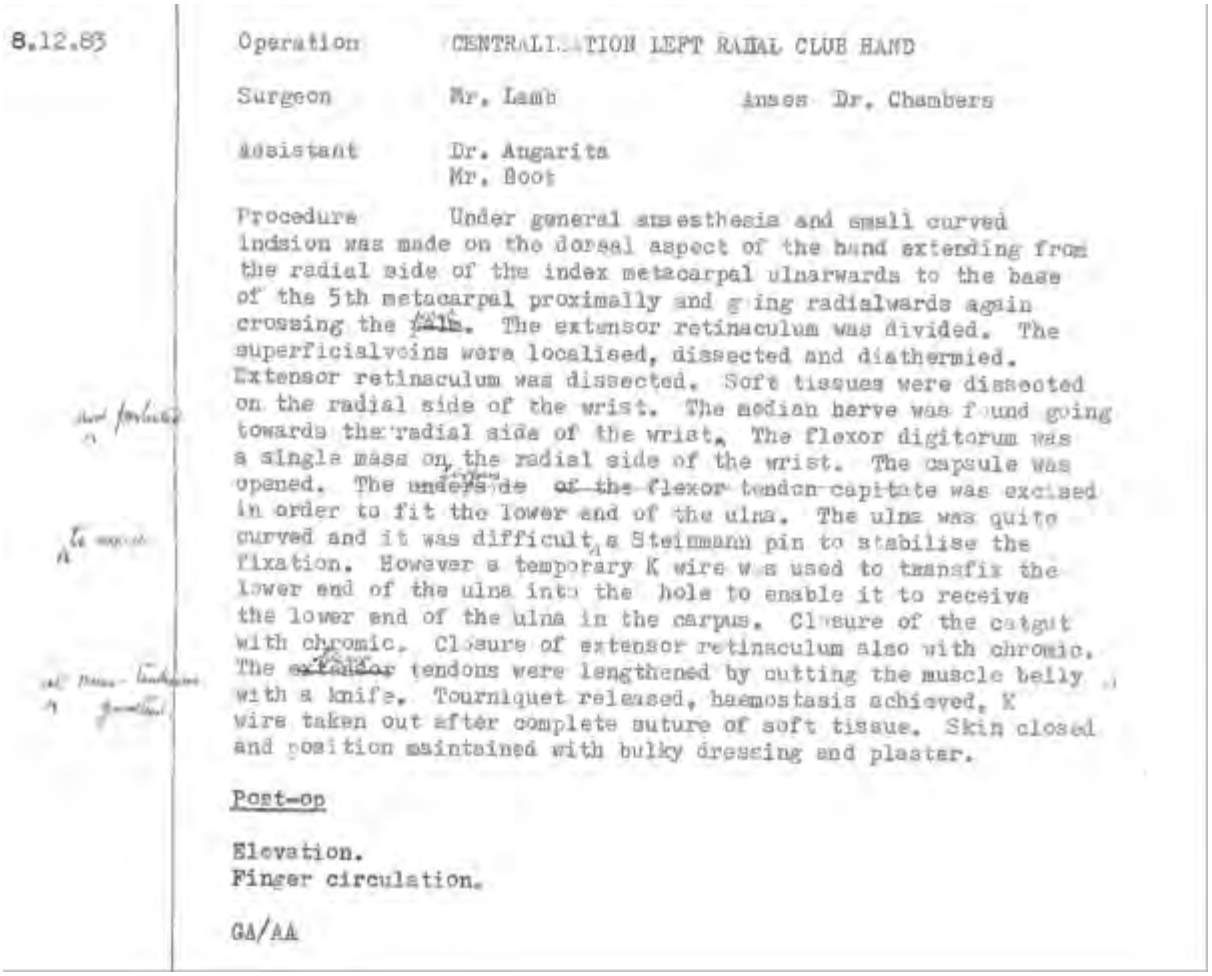
On the right side she has hypoplasia of the radius with with relatively mild wrist deformity. For the first time has had a corrective night splint which she finds uncomfortable. In actual fact the wrist can almost be corrected actively without She has most remarkably good finger function with no contractures or restrictions on active movement. She has a good thumb with active IP flexion/extension but no true thenar muscle function. She has contractured web.
The possibilities here are shortening of the ulna or lengthening of the radius. The function in this hand is so good that it may well be a case where relatively little lengthening of the radius is required and would be justifiable. The thumb would then be helped by deepening of the thumb web and opposition transfer. She appears to have normal tendon function in the ring finger.

On the left side she has a short forearm with absence of radius. The club hand deformity is not correctable due to soft tissue contractures. She has stiff MP joints with limited flexion of about 20 deg. Each PIPJ has a flexion contracture more marked on the radial side. There is a very poor profundus tendon function. There is a poorly developed thumb which todate they have wished to be retained. I think there would be a reasonable indication for centralisation but I have warned the parents it is not lilely to improve function nor would it be to her detriment.

LT Mr Bolton cc GP.
DWL/MN

Figure 28: Clinical notes for the above patient with bilateral radial dysplasia.

CASE 18 (continued): *Radial Longitudinal Dysplasia*



8.12.83

Operation CENTRALI..TION LEFT RADIAL CLUB HAND

Surgeon Mr. Lamb Anaes Dr. Chambers

Assistant Dr. Angarita
Mr. Boot

Procedure Under general anaesthesia and small curved incision was made on the dorsal aspect of the hand extending from the radial side of the index metacarpal ulnarwards to the base of the 5th metacarpal proximally and going radialwards again crossing the ~~pain~~. The extensor retinaculum was divided. The superficial veins were localised, dissected and diathermied. Extensor retinaculum was dissected. Soft tissues were dissected on the radial side of the wrist. The median nerve was found going towards the radial side of the wrist. The flexor digitorum was a single mass on the radial side of the wrist. The capsule was opened. The ~~undersi de~~ ~~of the~~ flexor tendon capitate was excised in order to fit the lower end of the ulna. The ulna was quite curved and it was difficult a Steinmann pin to stabilise the fixation. However a temporary K wire was used to transfix the lower end of the ulna into the hole to enable it to receive the lower end of the ulna in the carpus. Closure of the catgut with chromic. Closure of extensor retinaculum also with chromic. The ~~extensor~~ tendons were lengthened by cutting the muscle belly with a knife. Tourniquet released, haemostasis achieved, K wire taken out after complete suture of soft tissue. Skin closed and position maintained with bulky dressing and plaster.

Post-op

Elevation.
Finger circulation.

GA/AA

Figure 29: Surgery notes for left centralisation.

CASE 19: *Radial Longitudinal Dysplasia*

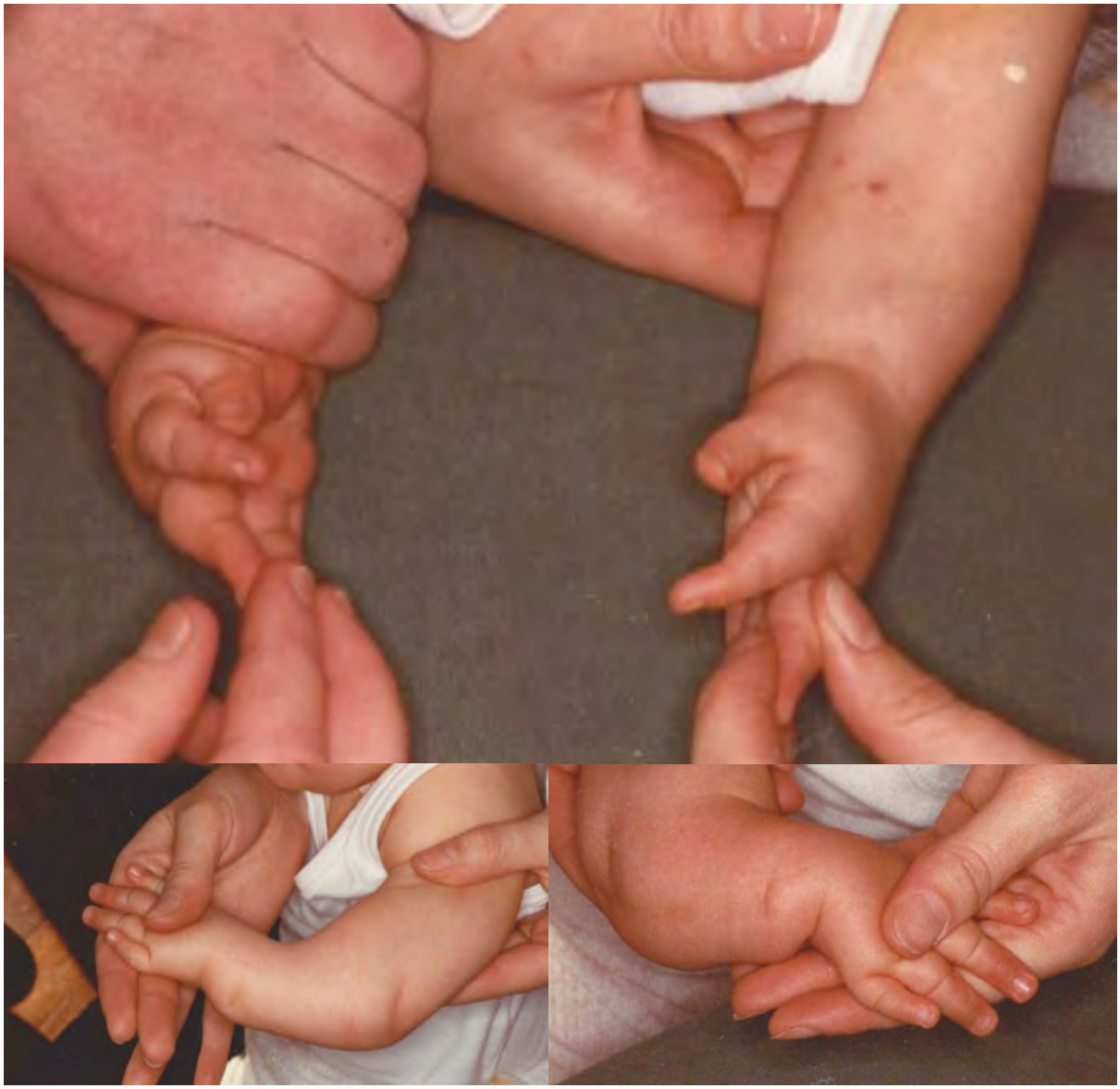


Figure 30: This is a child with TAR syndrome – Thrombocytopenia with aplasia of the radius, who presented with bilateral radial dysplasia. Note the thumbs that are present bilaterally, as is usually the case in this condition. He underwent bilateral centralizations.

CASE 19 (continued): *Radial Longitudinal Dysplasia*

CLINICAL HISTORY—*Continuation Sheet*

DATE

28. 2. 85

Pre-operative diagnosis.

TAR syndrome (thrombocytopenia) absent radius for centralisation of the right radius.

OPERATION: Centralisation of the right radius.

Surgeon: Mr. Lamb — Anaes. Dr. Chambers
Assistant: Mr. Batten — Anaesthetic general
Mr. Luk

Tourniquet time 2 hours and 4 minutes.

The patient was placed under a general anaesthetic. The platelets provided were then given by a left foot.. After they had been in for a few minutes the right arm was elevated, exsanguinated and a tourniquet applied to the upper arm. The area was then prepped and draped in the usual fashion using Savlon. A sinusoidal incision was made over the dorsal and radial aspect of the wrist from the mid dorsum of the hand to the middle of the forearm on the volar aspect. This incision was deepened preserving the superficial vein and through the deep fascia into the muscle compartment. The median nerve was then exposed and protected throughout the operation. A muscle belly assumed to be brachioradialis was then detached from the carpus and held with a cocka. The extensor tendons were then clearly displayed after releasing of the retinaculum and the carpus displayed at its most proximal end. The central carpus including the lunate and a very small amount of the scaphoid was then excised to make a small notch in which the ulna could be placed. The ulna was then dissected at its distal end and soft tissue release was then carried out to enable the ulna to sit comfortably in the notch made in the proximal row of the carpus. This was then stabilised by a retroigraded .45 mm. K wire along the intramedullary aspect of the shaft. X-ray showed this to be in an adequate position, the brachio-radialis tendon was then attached to the dorsal side of the carpus and the retinaculum was then closed with interrupted catgut and using 20 chromic catgut and interrupted suturing the two carpal buttresses were closed against the ulna. The tourniquet was then released and haemastasis was achieved This rapid return of effusion to the distal rim. The subcutaneous layer was closed with 30 catgut and the skin closed with 30 catgut. A wool and crepe bandage was applied and an above elbow dressing with a supporting dorsal back slab was applied for support.

Post-op management. The patient should be kept elevated . A very close watch should be kept on her bleeding and swelling. There is platelets available for excessive bleeding.

JB/IJR

Figure 31: Surgery notes for right centralisation.

CASE 20: *Radial Longitudinal Dysplasia*

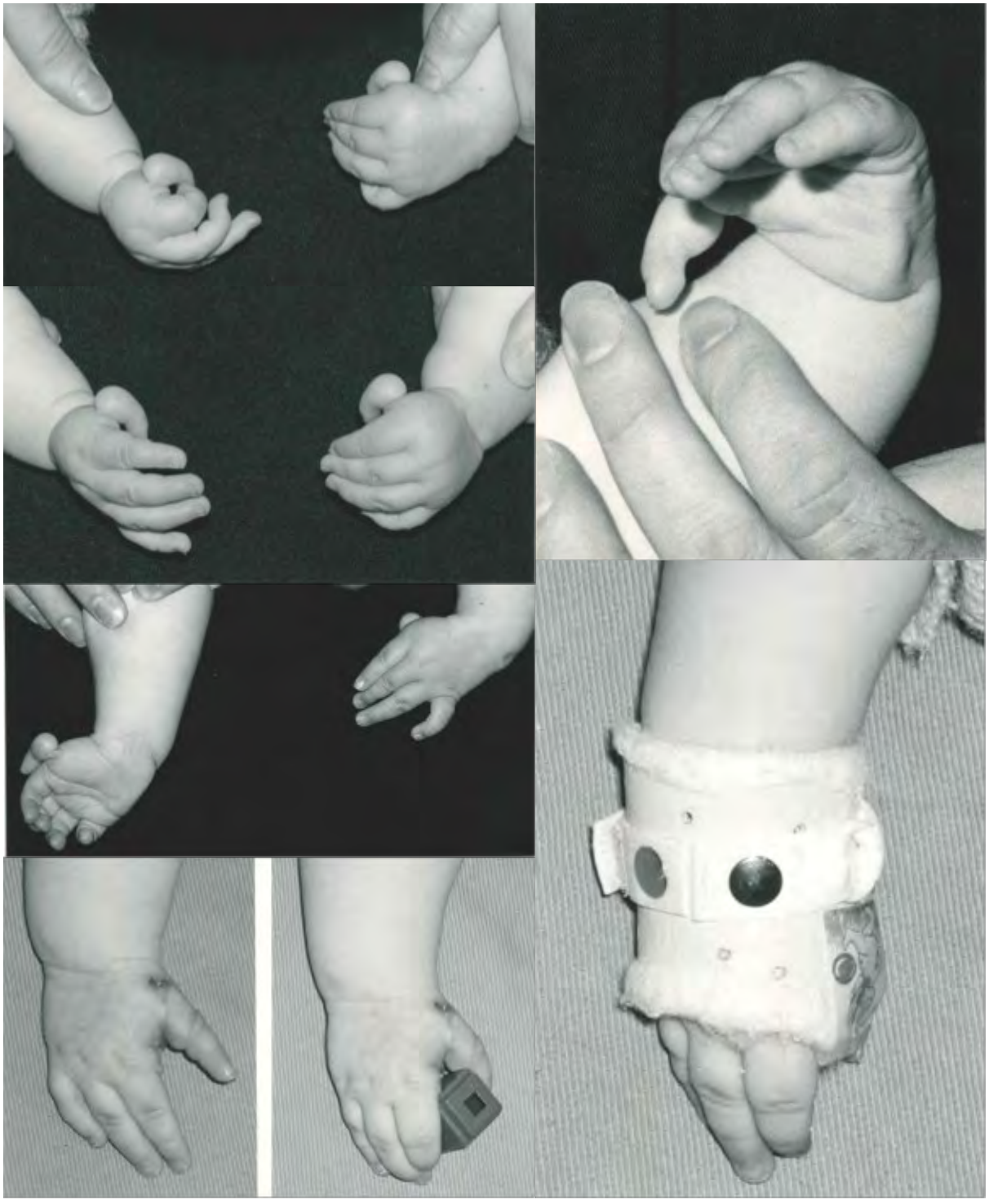


Figure 32: This child presented with bilateral radial dysplasia. The left wrist did not undergo centralisation. On the right hand, the child has a Type 4 hypoplastic thumb and he underwent a pollicisation.

CASE 20 (continued): *Radial Longitudinal Dysplasia*

CLINICAL HISTORY

31.1.86

Seen at clinic by mr Lamb

DIAGNOSIS : ? FANCONI SYNDROME

HISTORY :

This little boy of three months has bilat, hypoplasia of the radius, more makred on the left side. There is no obvious deformity on the right side but some radial deviaiton on the left side which is easily correctable. There is bilat, dangle thumbs. I have explianed that these are functionless and will not grow and do not justify any attempts at surgical reconstruction. He has got good four fingered hand with excellent movement and should get good result from pollicisation.

We will see in four months and start working out the details of this and in the meantime Helen will try and show the parents a child who has had had pollicisaiton or show them a film.

Parents think the child ahs some difficulty in hearing and they should consult Dr Stark about this.

LT Mr Batchelor cc Dr Stark cc GP.
DWL/MW

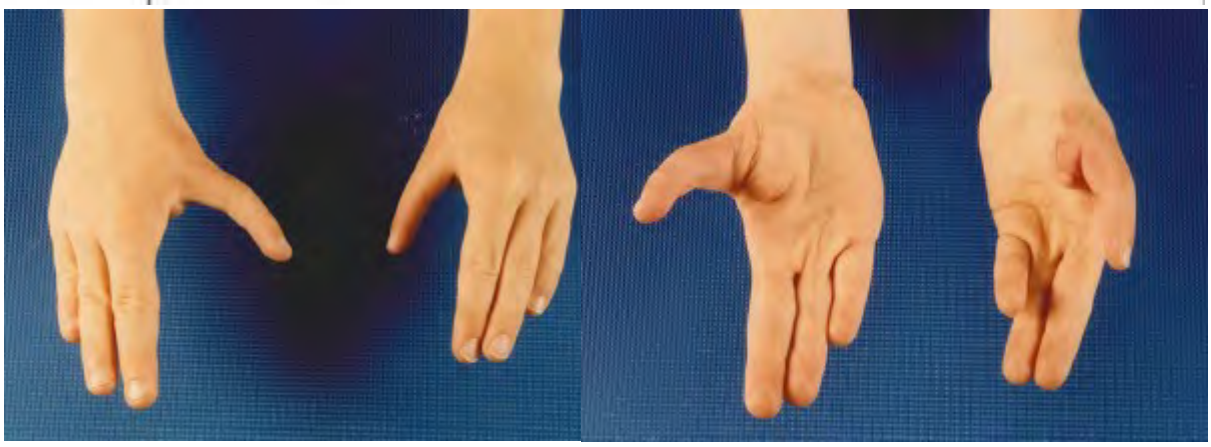


Figure 33: Clinical notes of the above patients. Decision was made to pollicise both index fingers.

Recent Developments in Radial Dysplasia Surgery and Pollicisation

THE NAME 'CENTRALISATION' is perhaps synonymous with Douglas Lamb's method of fixing the carpus over the ulna and aligning the wrist with a K wire inserted from the ulnar diaphysis into the third metacarpal. Although described in the 1970s, the procedure is still popularly performed, especially for patients who present late and for recurrent cases. In 1985, Buck-Gramcko of Germany described the procedure of radialisation, which does not involve the creation of a carpal slot but simply balancing the carpus over the ulna. (Buck-Gramcko, 1985) In radialisation, a K-wire is inserted in line with the second metacarpal instead of the third. There is also a stronger emphasis on tendon transfer. It is generally felt that radialisation may be a more attractive option as it is less destructive to the carpus, but the procedure is probably more suitable for younger children without fixed deformities.

Whether centralisation or radialisation is chosen, there is always the concern that undue stress would be placed on the ulnar physis when positioning the carpus, and therefore affect growth potential of the forearm. These concerns ushered in the era of pre-operative distraction, which was not used routinely back then. Nowadays, it is well established that distracting the wrist will lead to effective lengthening of soft tissues on the radial side and easier positioning of the carpus on the ulna, with reduced stress on the growth physis. However, it is unproven that prior distraction will lead to a lower rate of recurrence, even though growth of the forearm is probably better preserved.

In terms of pollicisation, much of the debate nowadays focused on the incisions used and whether the intrinsics should be reconstructed or not. Some of the more popular eponymous incisions used nowadays include the 'Buck-Gramcko', 'Foucher' or the 'Dallas' incisions. Douglas never specified an eponymous name for his incision, simply describing it as one that was made "circumferentially at the level of the metacarpal phalangeal joint of the index finger and taken dorsally". In terms of intrinsic reconstruction, he described: "the interosseous muscles were then reinserted, woven through the extensor tendon between the metacarpal phalangeal interphalangeal joint". It would appear that his preference was to reconstruct the intrinsics, although there is currently a small group of surgeons who would recommend that intrinsic reconstruction is not necessary.

As mentioned, his preference would be to pollicise the index finger wherever possible, regardless of unilateral or bilateral anomalies. In contrast, there is probably an increasing reluctance to perform pollicisation in patients with radial dysplasia, as the index finger is often stiff and therefore the result is a 'poor thumb'. Furthermore, there is always the concern that the straightened wrist would deviate again, causing the child to change from a radial to ulnar prehension, thereby neglecting the pollicised digit in day to day activities.

I-A-2-ii. Ulnar Longitudinal Deficiency

ULNAR LONGITUDINAL DEFICIENCY is a lot less common than its radial counterpart. The diagnosis of ulnar longitudinal deficiency remains challenging, and the condition probably presents as a spectrum from a type of phocomelia ***(see above)*** to a shortened forearm only (affecting both radius and ulna) or to an upper limb with normal humerus and forearm but missing digits distally (ulnar longitudinal deficiency of the hand plate only, 1-B-2-ii). Despite being an 'ulnar' deficiency, the thumb can be affected as well, adding to the confusion.

CASE 21: *Ulnar Longitudinal Dysplasia*

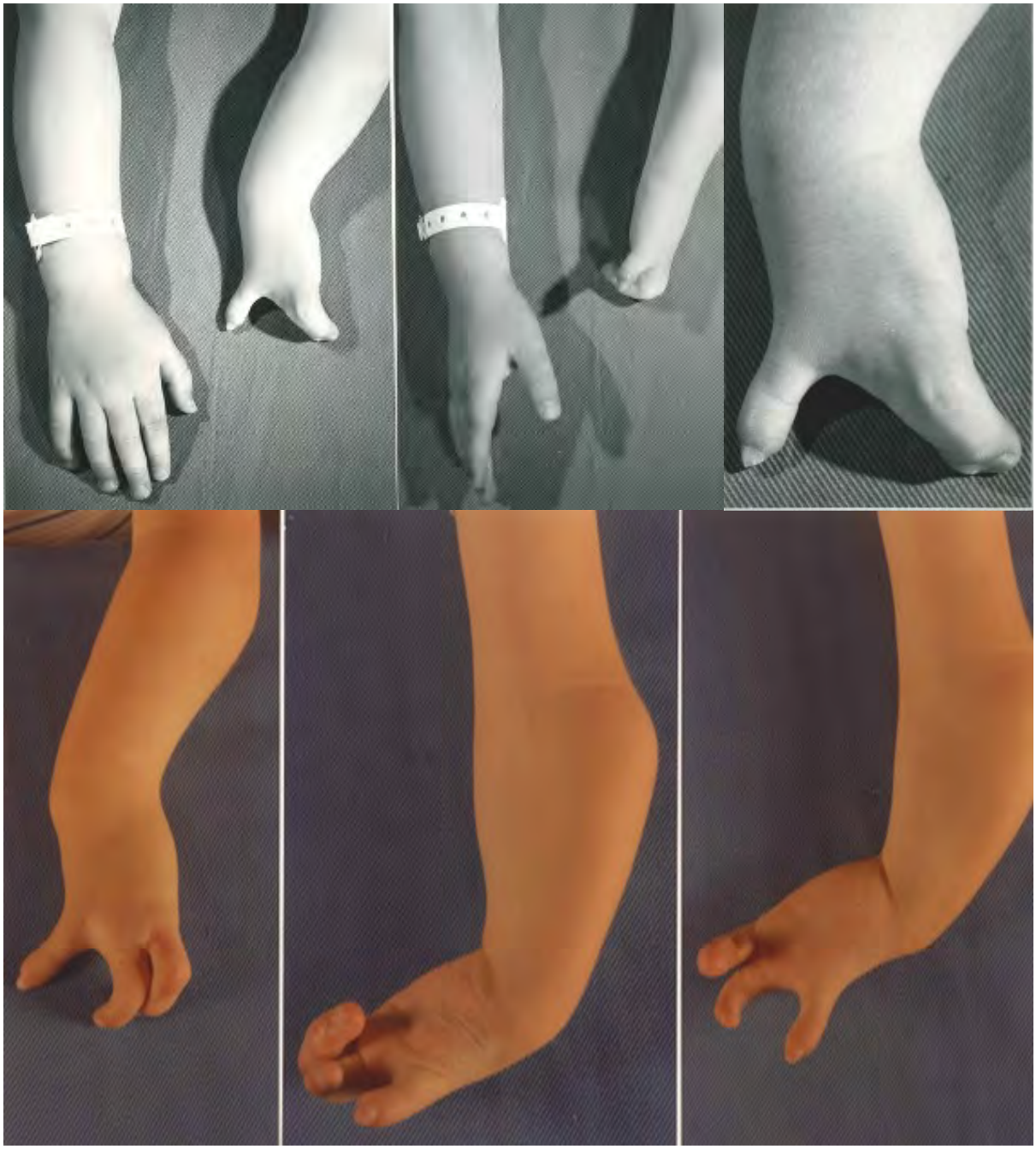


Figure 34: Original diagnosis (DWL): *'congenital absence of the left ulna.'* This patient has a left sided ulnar dysplasia affecting the entire upper limb. The humerus appear to be of equal length to the right but the diagnosis is made by observation of a short forearm and missing or hypoplastic digits distally. In this case, the patient has a hand with syndactylised 2 digits and a hypoplastic thumb.

CASE 21 (continued): *Ulnar Longitudinal Dysplasia*

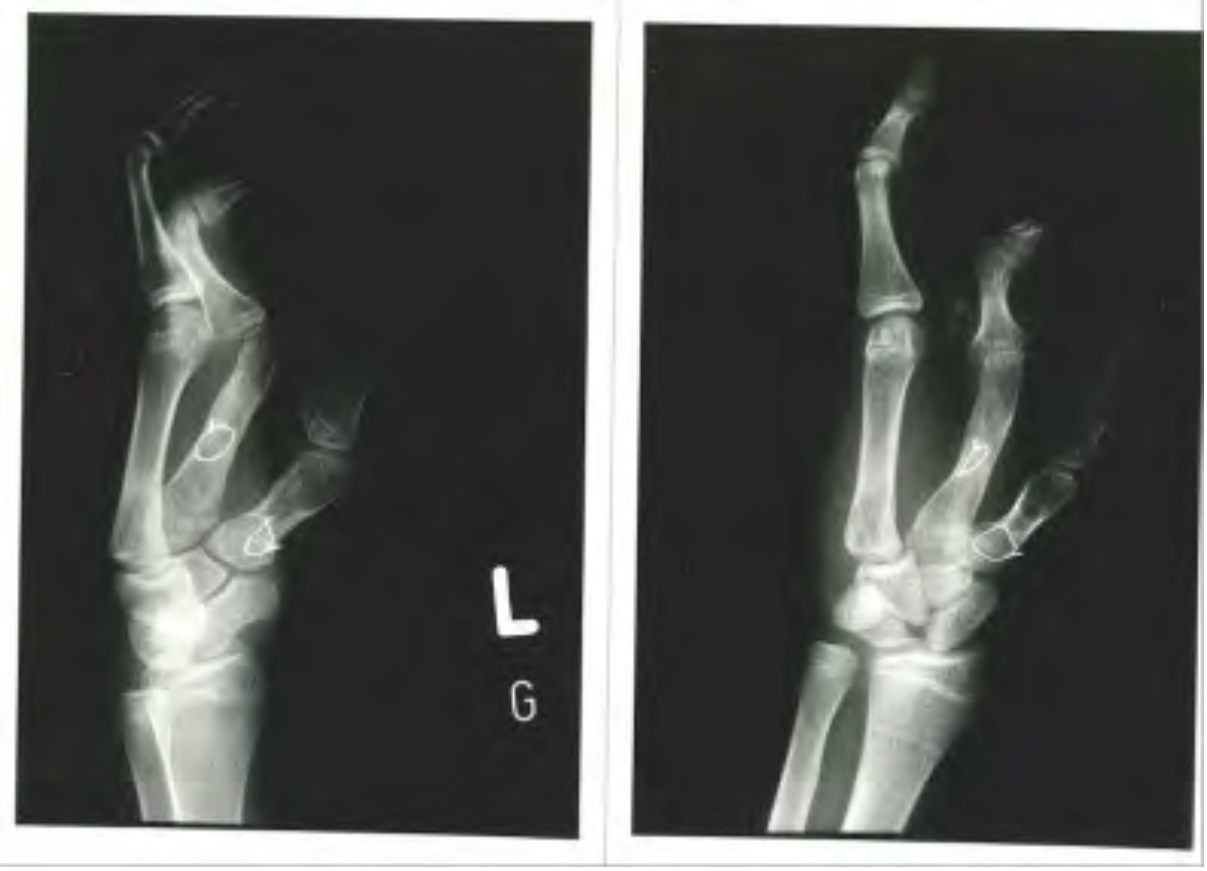


Figure 35: Radiograph of left sided ulnar dysplasia for above patient.

CASE 21 (continued): *Ulnar Longitudinal Dysplasia*

15.8.85

Operation SEPARATION OF THUMB AND IDNEX FINGERS FROM BONY JUNCTION AT RADIO METACARPAL

Surgeon Mr. Lamb

Anaes Dr. Howie
TT 58 mins

Assistants Dr. Hoffman
Dr. Skak

Procedure Under adequate general anaesthesia and pre-tested well padded tourniquet control the z plasty incisions were marked out on the thumb web space. Flaps were raised and carefully dissected. The thumb metacarpal was then visualised dorsally and the extensor tendon to thumb and index finger identified and joined proximally over the metacarpal. The metacrpal was then freed subperiosteally out to the mnetacarpal head and the proximal phalanges to index and thumb were identified. The metacarpal head was then divided in its centre and the division of the metacarpal progressed proximally and ulnarly with the bulk of the metacarpal remaining with thumb.

Exposure of the radial side of the ulnar of the two metacarpals was then done and the dorsal interosseous freed. Sub-periosteal dissection was made on the middle and distal aspect of the metacarpal and soft tissue was freed between the ulnar part of the separated metacarpal and the persistent third metacarpal. Kirschner wire was used to fix the index proximal phalanx to middle phalanx and the metacarpal head and distal shaft of the separated thumb metacarpal to the third metacarpal shaft. Dorsal interosseous was then sutured with interrupted catgut to the long extensor tendon to index finger. There was a good passive motion of the thumb metacarpal at this stage.

Tourniquet was then deflated. Haemostasis was obtained. The Kirschner wires were cut off just outside the bony level and the z plasty was closed with interrupted catgut sutures. Sterile dressing of sofratulle, blue gauze, fluff and a plaster slab applied.

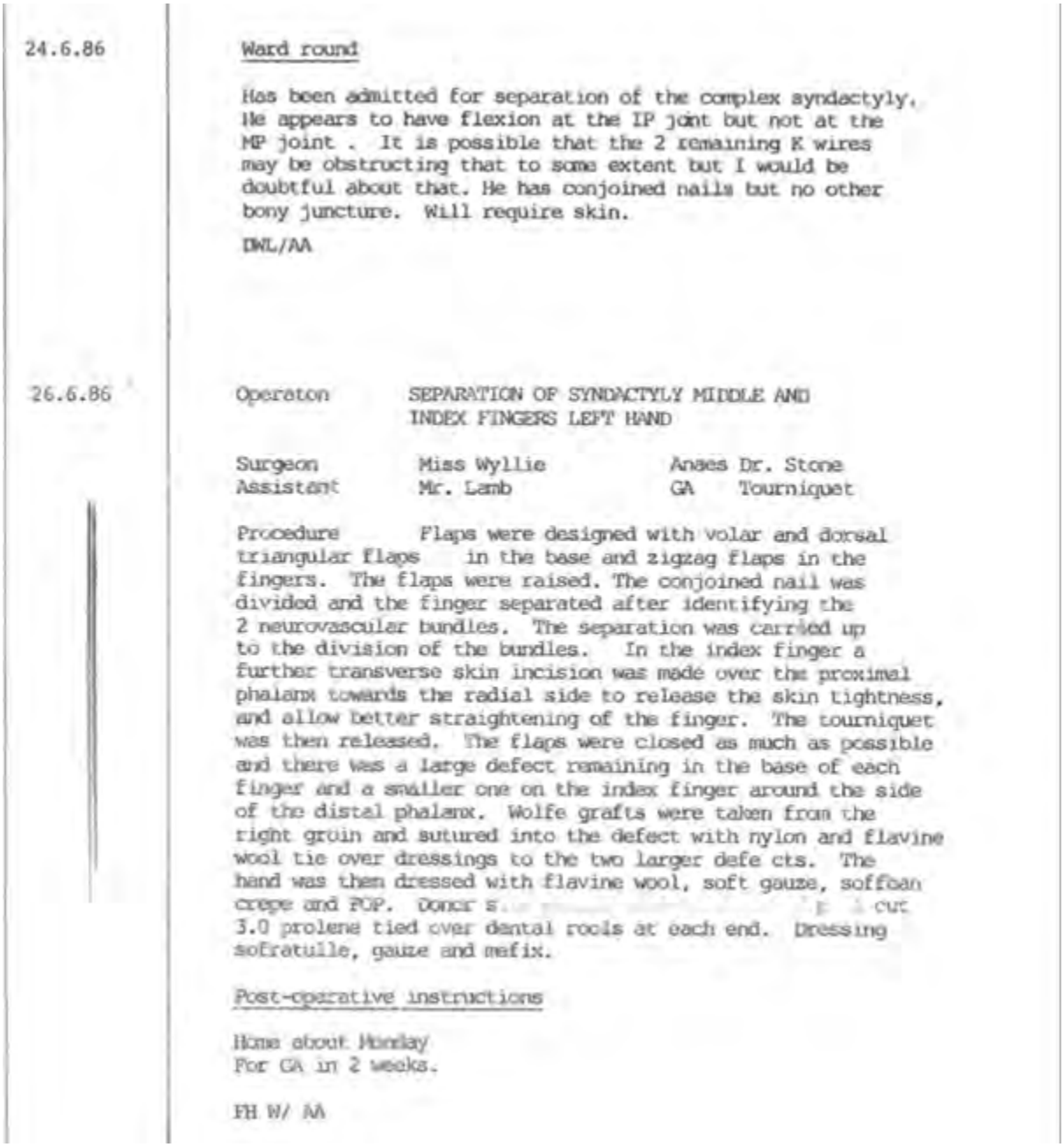
Post-operative instructions

Full time elevation.
Patient to be discharged in 48 hours.
See in the clinic in 2 weeks.

RPH/AA

Figure 36: Operative management of this patient included separation of the syndactyly, but nothing else was done to the forearm.

CASE 21 (continued): *Ulnar Longitudinal Dysplasia*



24.6.86

Ward round

Has been admitted for separation of the complex syndactyly. He appears to have flexion at the IP joint but not at the MP joint . It is possible that the 2 remaining K wires may be obstructing that to some extent but I would be doubtful about that. He has conjoined nails but no other bony juncture. Will require skin.

DWL/AA

26.6.86

Operaton SEPARATION OF SYNDACTYLY MIDDLE AND INDEX FINGERS LEFT HAND

Surgeon Miss Wyllie Anaes Dr. Stone
Assistant Mr. Lamb GA Tourniquet

Procedure Flaps were designed with volar and dorsal triangular flaps in the base and zigzag flaps in the fingers. The flaps were raised. The conjoined nail was divided and the finger separated after identifying the 2 neurovascular bundles. The separation was carried up to the division of the bundles. In the index finger a further transverse skin incision was made over the proximal phalanx towards the radial side to release the skin tightness, and allow better straightening of the finger. The tourniquet was then released. The flaps were closed as much as possible and there was a large defect remaining in the base of each finger and a smaller one on the index finger around the side of the distal phalanx. Wolfe grafts were taken from the right groin and sutured into the defect with nylon and flavine wool tie over dressings to the two larger defe cts. The hand was then dressed with flavine wool, soft gauze, soffban crepe and POP. Donor s cut 3.0 prolene tied over dental rools at each end. Dressing sofratulle, gauze and mefix.

Post-operative instructions

Home about Monday
For GA in 2 weeks.

FH W/ AA

Figure 36 (continued): Operative management of this patient included separation of the syndactyly, but nothing else was done to the forearm.

CASE 22: *Ulnar Longitudinal Dysplasia*

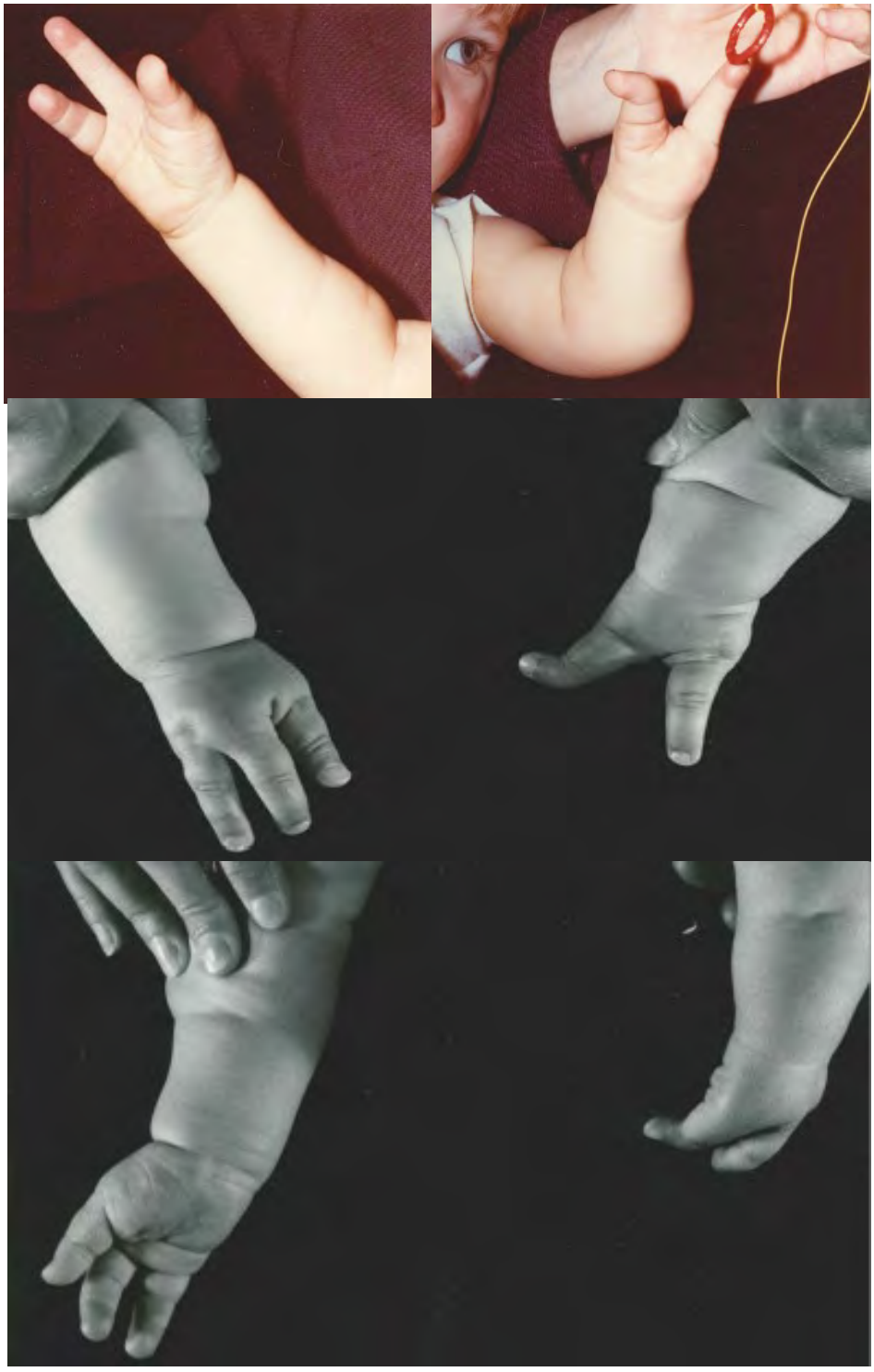


Figure 37: The patient was diagnosed with bilateral ulnar dysplasia. Most of these diagnoses were described quite accurately with congenital absence of the ulna. Management of this patient included removal of the delta phalanx of the left thumb.

CASE 22 (continued): *Ulnar Longitudinal Dysplasia*

30.8.74

Seen at Friday Clinic by Mr. Lamb

This little baby was seen in the Simpson by Mr. Watson who has referred the child here. The child is three weeks old. Condition seems tobe satisfactory otherwise.

The problems are in the upper limbs.

The right hand has two digits and a thumb. The thumb is lying in the same plane as the digits but he is already trying to bring it round into opposition. There may be someproblems with contracture but we will have to wait and see. I would anticipate that this would be the only = possible surgical treatment that might be required.

On the left side, there is a short forearm with a thick curved radius and a short ulna. There is some hyper-extension of the elbow. There are two digits. There is slight lobster claw effect but he is able to bring the two digits together. The more ulnar of these digits is showing some signs of angulation and there may be a delta phalanx. There would certainly be no justification for surgery here at the moment. We should simply watch and see.
=
The child was x-rayed three weeks ago by Mr. Watson, and there is no justification forfurther x-ray.
Clinical photography was not possible today.

Letter to Mr. Watson cc to Dr. Irons. DWL/PMB.

14.10.76

OPERATION : Removal of delta-phalanx left thumb.

SURGEON : Mr. D.W. Lamb.
ASSISTANT : Mr. C.B. Clowes. ANAES : Dr. R. Park

PROCEDURE :

Through an 'S' shaped in cision centred over the dorsum of the delta-phalanx, a flap of capsule and periosteum was raised on the ulnar side and the delta-phalanx removed. Unfortunately the capsular flap became detached and was used as a free graft.
Skin closed with interrupted nylon and a wool bandage with plaster back slab was applied holding thumb in the corrected position.

CBC/PMB.

Figure 38: Operation notes for the above patient with bilateral ulnar dysplasia

CASE 23: *Ulnar Longitudinal Dysplasia*

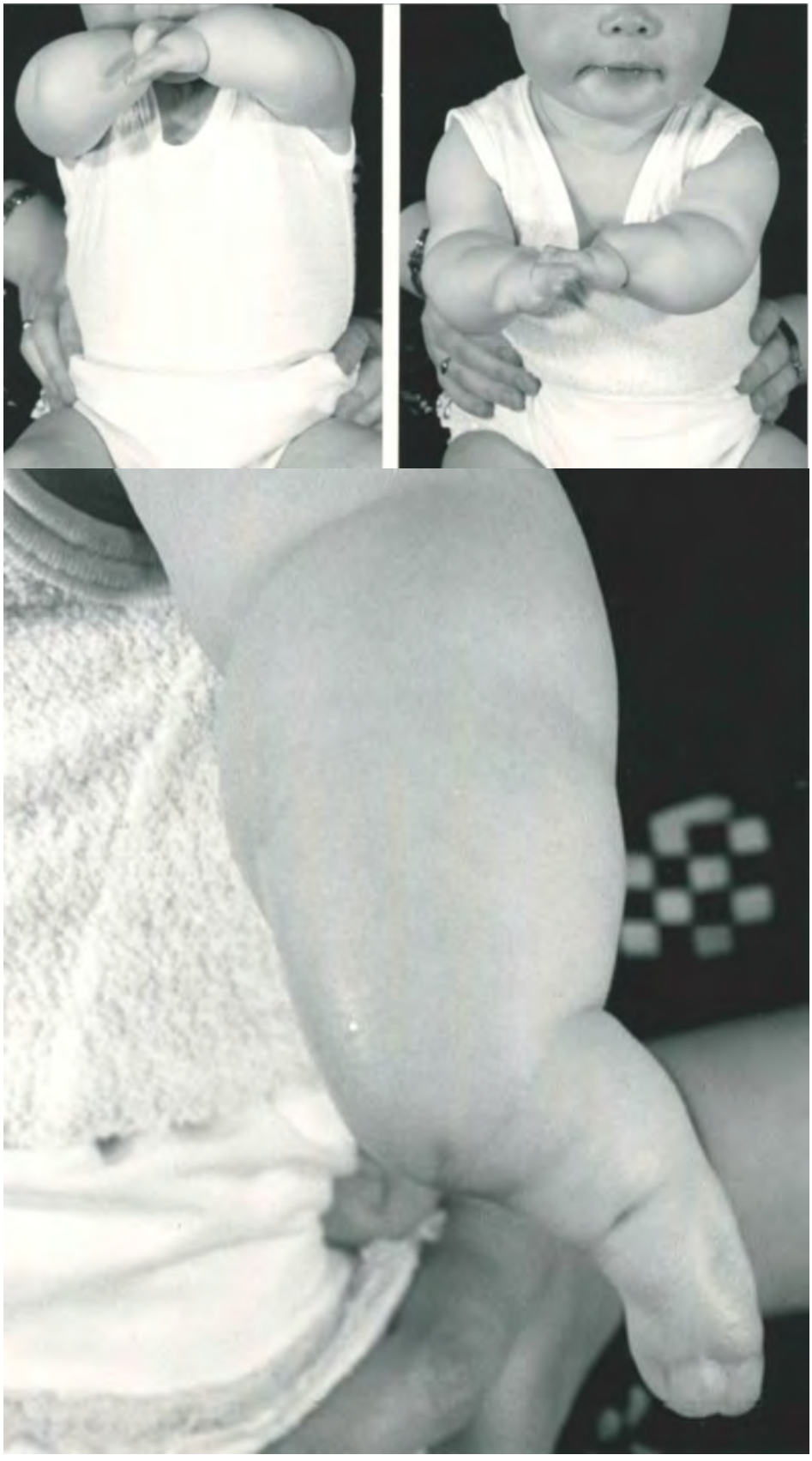


Figure 39: Original diagnosis (DWL): *'congenital absence of the ulna bilaterally.'* The possibility of toe transfer was discussed but the child did not undergo any surgery as she was making excellent use of her upper limbs.

CASE 24: *Ulnar Longitudinal Dysplasia*

Figure 40: Original diagnosis (DWL): *'congenital absence of the left ulna'*. This patient has left ulnar dysplasia affecting the entire upper limb. The two digits were syndactylised and separated. No further surgery was performed on the forearm.

CASE 25: *Ulnar Longitudinal Dysplasia*

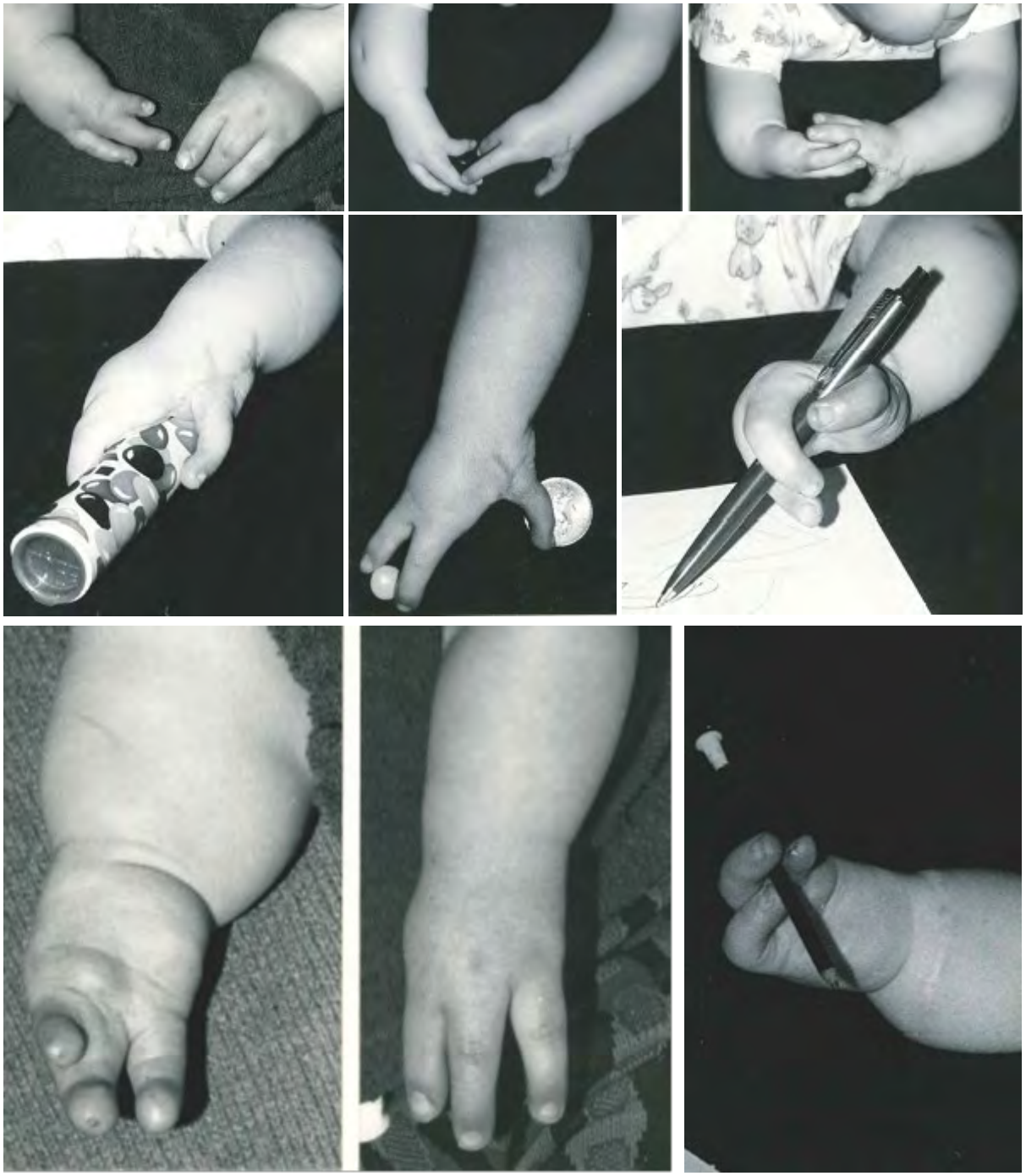


Figure 41: Original diagnosis: (DWL) '*bilateral congenital absence of the ulna?*' On the left, this patient has an absent thumb but to improve prehension, the child underwent pollicisation of the little finger. Ulnar or little finger pollicisation is an unusual procedure that is carried out if the child has an absent thumb and a predominantly ulnar prehension.

CASE 25 (continued): *Ulnar Longitudinal Dysplasia*

Figure 42: Post-op photographs of the above patient with bilateral ulnar longitudinal dysplasia.

IB. MALFORMATION OF THE HAND PLATE:
ABNORMAL AXIS DIFFERENTIATION (LATE LIMB PATTERNING)

I-B-1. Malformation of the Hand Plate:
Proximal-distal Axis

THE DIVISION of malformations in the OMT classification into these two main groups (entire upper limb and hand plate) denotes that an insult to the developing limb happened at a later stage. In the 2020 OMT update, Goldfarb et al. (2020) assigned the words 'early patterning' to malformations of the entire upper limb and 'late patterning' to malformations of the hand plate.

One would expect that in malformations of the hand plate that the forearm would generally be normal. However, this is not always the case as several hand conditions may also affect the forearm because these are longitudinal deficiencies. The subclassifications of malformations of the hand plate are similar to that of the entire upper limb.

Similar to malformations of the entire upper limb, hand plate malformations of the proximal-distal axis result from predominant defects in the apical ectodermal ridge. This gives rise to a variety of conditions distal to the wrist. For example, symbrachydactyly or transverse arrest affecting the hand plate results in shortened fingers, but with the forearm and wrist still present. Another condition is cleft hand, which has recently moved from an unspecified axis to the proximal-distal axis because of recent knowledge about its pathogenesis.

I-B-1-ii. Symbrachydactyly (with ectodermal elements)

SIMILAR TO symbrachydactyly of the entire upper limb, this condition presents with nubbins which are composed of ectodermal elements (nail, skin). This differentiates it from transverse arrest of the hand plate.

CASE 26: *Symbrachydactyly (Hand Plate)*

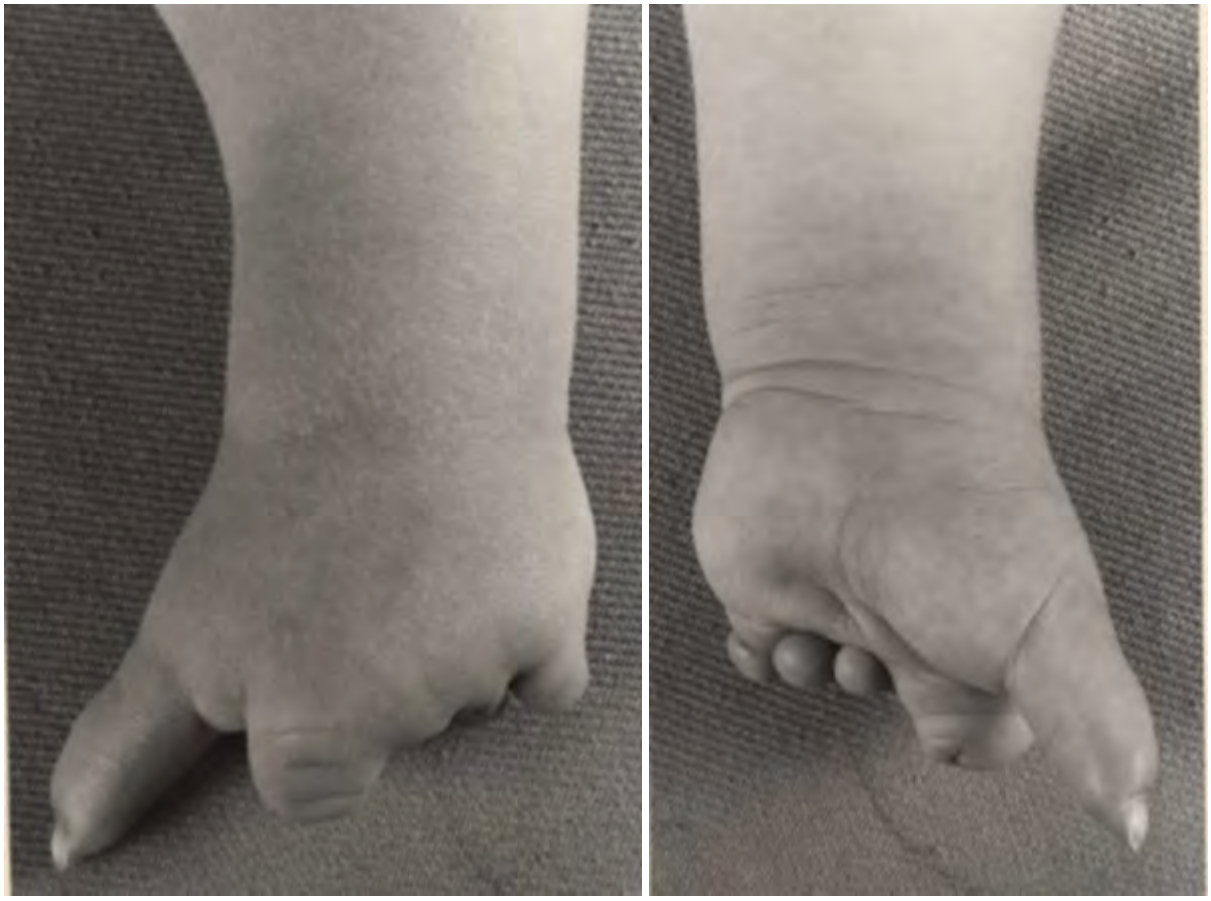
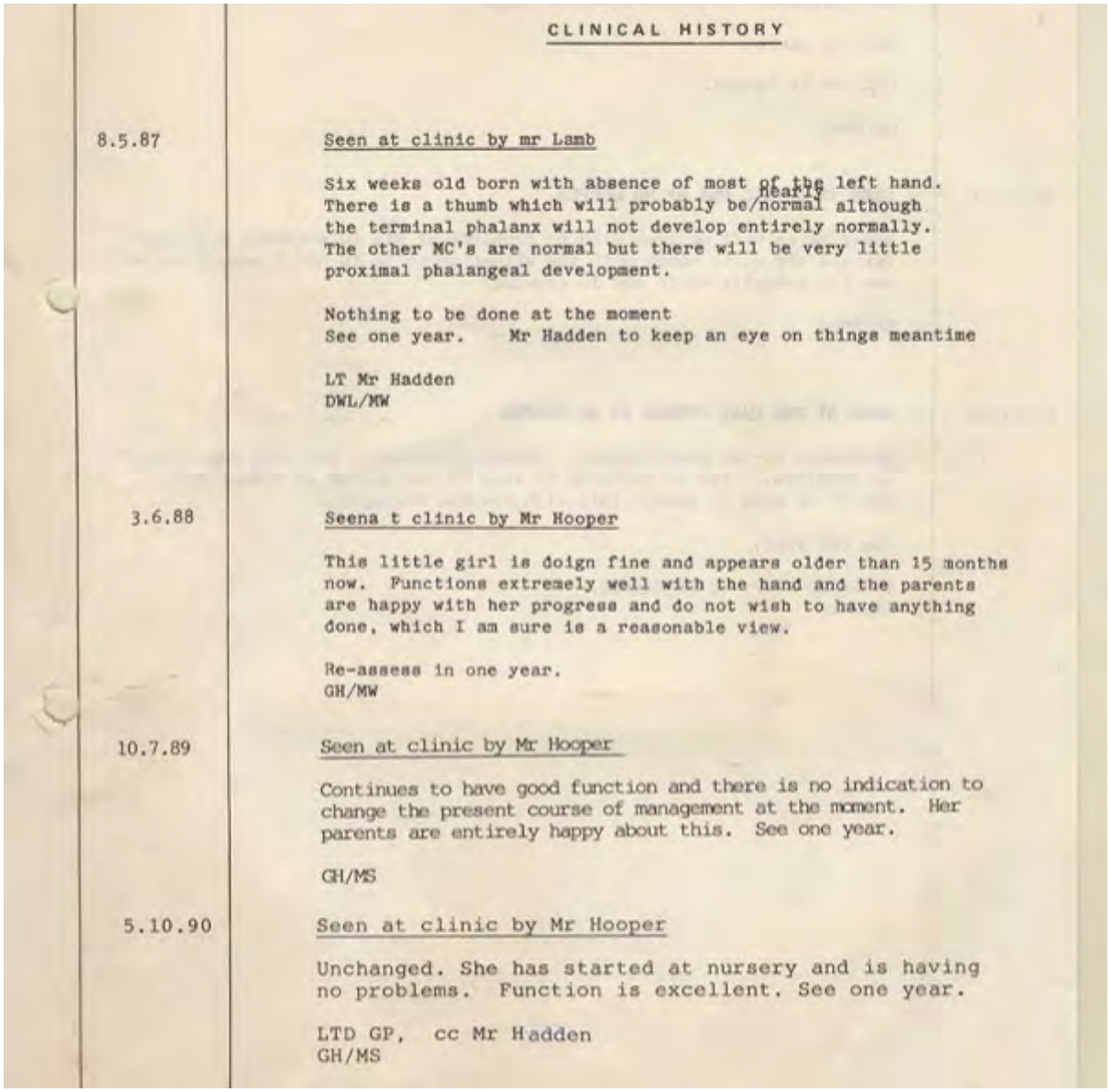


Figure 43: In this patient, you can see symbrachydactyly of the hand. The thumb is present with good thenar eminence. The index finger is shortened and the other digits consist of nubbins. This would be considered an oligodactylous or monodactylous type symbrachydactyly. In this patient, no further treatment was implemented. These children do remarkably well because of the good thumbs that they have.

CASE 26 (continued): *Symbrachydactyly (Hand Plate)*



CLINICAL HISTORY

8.5.87

Seen at clinic by mr Lamb

Six weeks old born with absence of most of the left hand. There is a thumb which will probably be/nearly normal although the terminal phalanx will not develop entirely normally. The other MC's are normal but there will be very little proximal phalangeal development.

Nothing to be done at the moment
See one year. Mr Hadden to keep an eye on things meantime

LT Mr Hadden
DWL/MW

3.6.88

Seena t clinic by Mr Hooper

This little girl is doign fine and appears older than 15 months now. Functions extremely well with the hand and the parents are happy with her progress and do not wish to have anything done, which I am sure is a reasonable view.

Re-assess in one year.
GH/MW

10.7.89

Seen at clinic by Mr Hooper

Continues to have good function and there is no indication to change the present course of management at the moment. Her parents are entirely happy about this. See one year.

GH/MS

5.10.90

Seen at clinic by Mr Hooper

Unchanged. She has started at nursery and is having no problems. Function is excellent. See one year.

LTD GP, cc Mr Hadden
GH/MS

Figure 44: Clinical notes for the above patient with symbrachydactyly of the hand.

CASE 27: *Symbrachydactyly (Hand Plate)*

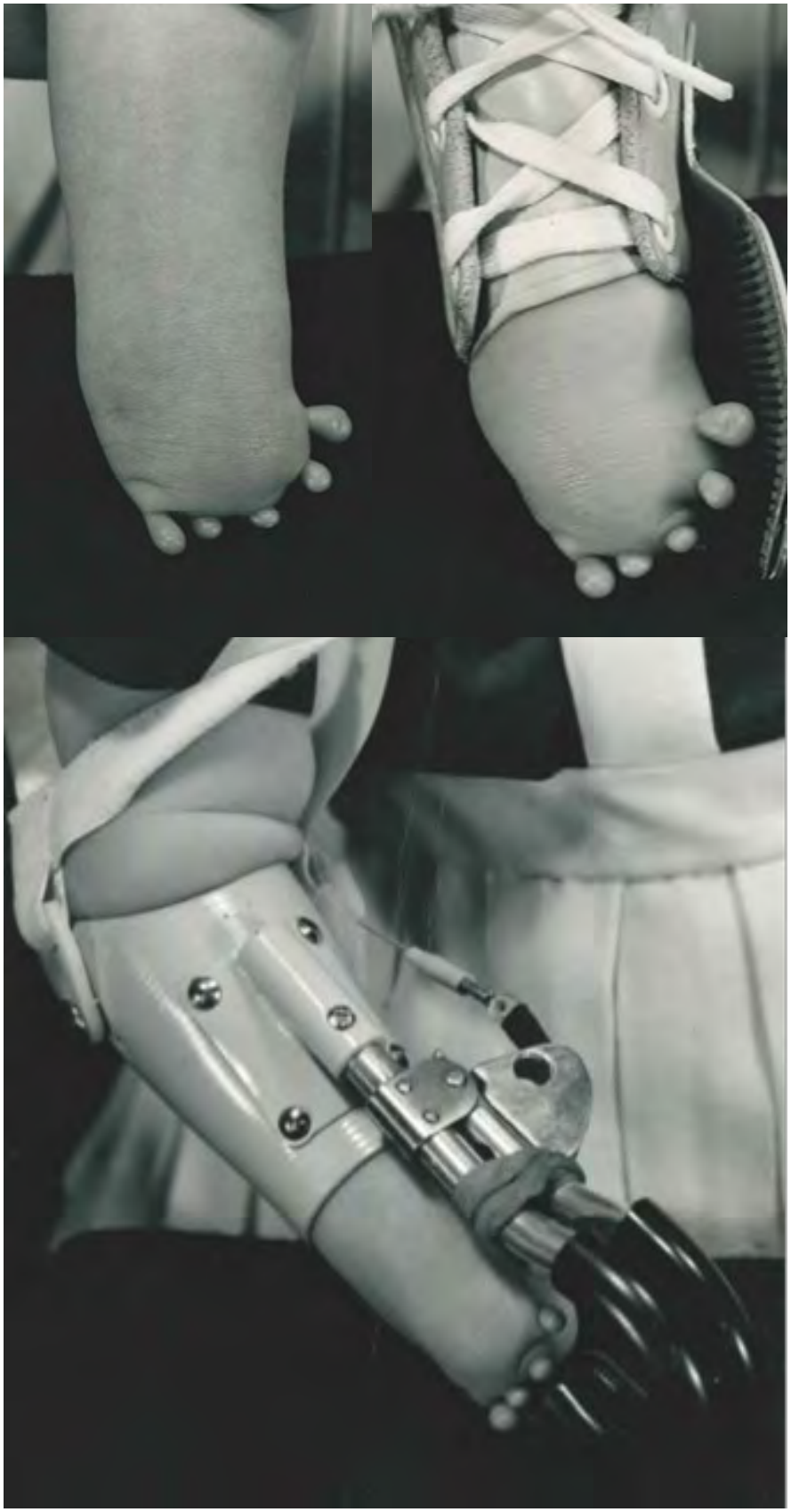


Figure 45: This is another case of hand plate syndactyly. You can see a wrist joint and several nubbins after that. In this patient, she was fitted with a specialist hook prosthesis after having her nubbins removed.

CASE 28: *Symbrachydactyly (Hand Plate)*

Figure 46: This is another case of right hand plate symbrachydactyly. Again you can see a wrist joint. The thumb is present but shortened. There are good thenar and hypothenar eminences. The digits are mainly represented by nubbins. This is another example of monodactylous symbrachydactyly. In this boy, the right hand was fitted with a cosmetic prosthesis.

CASE 29: *Symbrachydactyly (Hand Plate)*

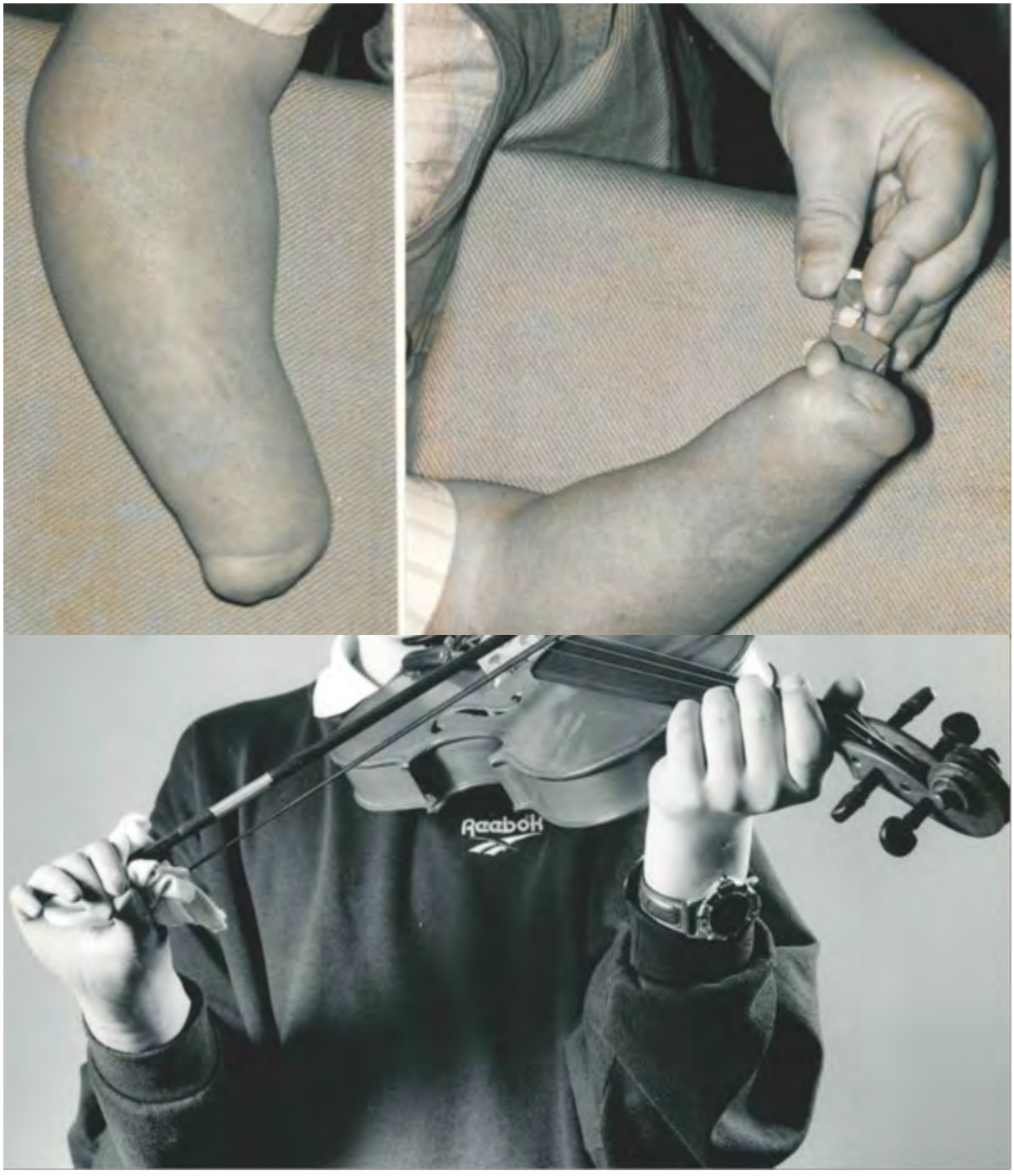


Figure 47: This is a severe symbrachydactyly affecting the junction of the entire upper limb and the hand plate. There appears to be a single digit arising from the radial aspect. He was fitted with a prosthesis and obviously makes very good use of it.

CASE 30: *Symbrachydactyly (Hand Plate)*

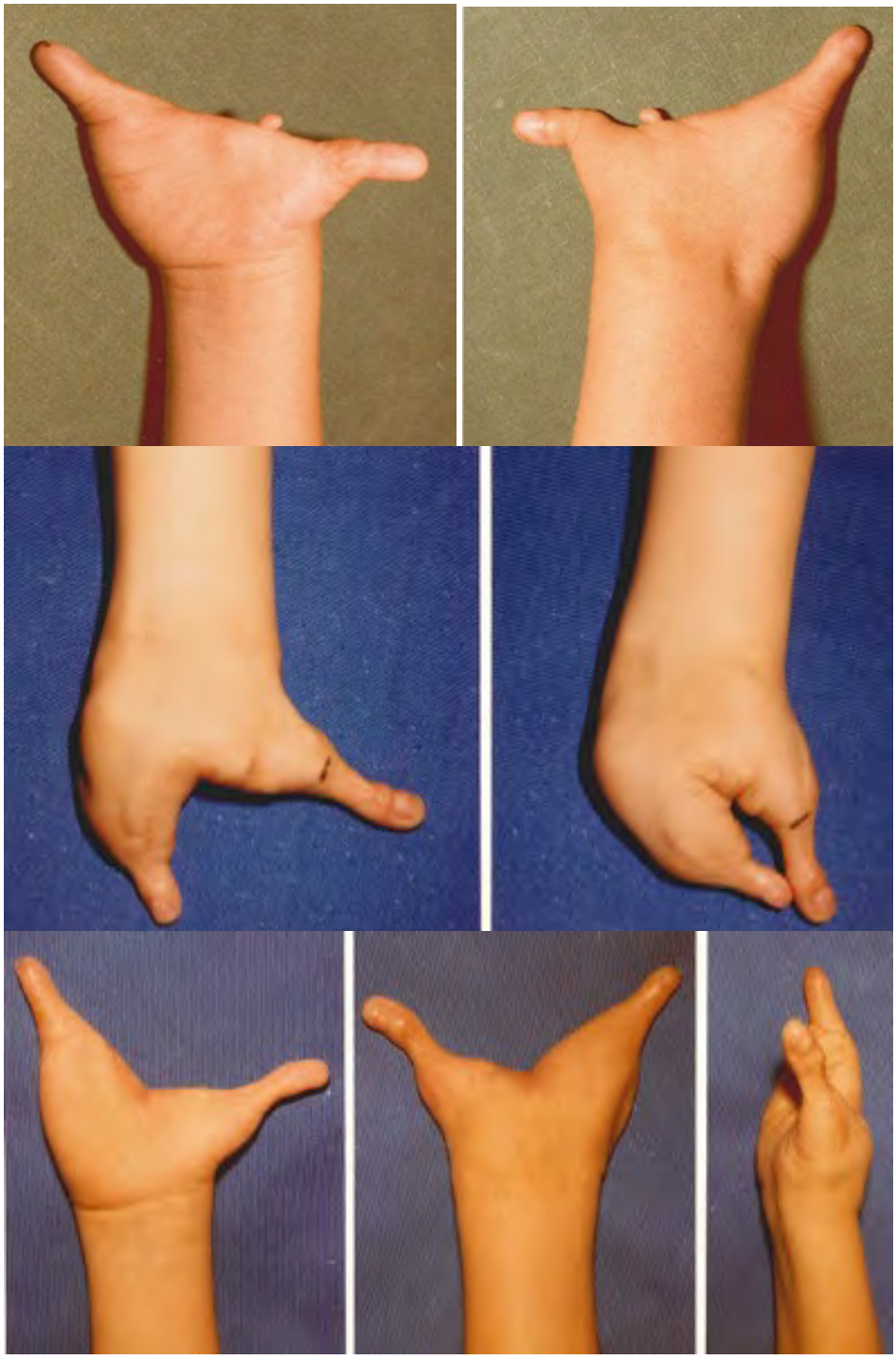


Figure 48: This patient was diagnosed as having a "*central defect*." The diagnosis is probably that of a symbrachydactyly rather than a cleft hand. She underwent transfer of the proximal phalanx from the left fourth toe to lengthen the thumb. This was a non-vascularised transfer. Subsequently she underwent a tendon transfer to improve the opposition of the thumb.

CASE 30 (continued): *Symbrachydactyly (Hand Plate)*

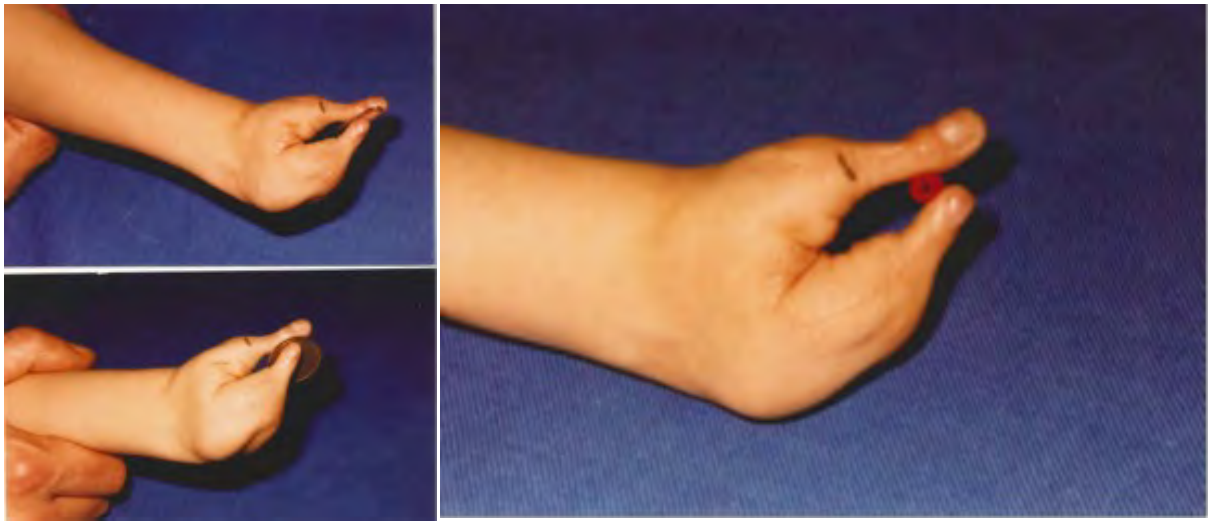


Figure 49: A display of function of the above patient.

29.9.78

Seen at clinic by Mr Lamb

DIAGNOSIS: Cong. abnormality (R) hand

HISTORY:

This little lad was referred by Mr Maisels from Liverpool. He has a central defect. Both ulnar and radial elements are not functional very effective. The thumb has deformed middle phalanx and floppy. The ulnar segment, on X-ray, has one bone phalanx but I think there is a little distal phalanx. Three is no active movemen in either segment. Passively the tip of the digits can be approximated. I am notcertain what the situation would be if the flexor tendon and superficialis tendon were present a graft elongation to try and provide active movement of the thumb segment may well be indicated. The ulnar segment requires to be osteotomised in a better positon.

I am not sure if Mr Maisels wishes to deal with the boy or if he would like to do so. The parents have agreed to come here.

L/T Mr Maisels cc GP
DWL/MW

Figure 50: Clinical notes for the above patient with symbrachydactyly of the hand rather than the original diagnosis of 'cleft hand'.

CASE 30 (continued): *Symbrachydactyly (Hand Plate)*

31.7.79

Seen on ward round by Mr. Lamb

This presents a difficult problem. Over past few months has developed some limited control of his thenar. The main stumbling block to the thumb component is the lack of middle phalanx which is floppy. I think if some bone stability could be obtained there this might help him. I am thinking of the possibility on transposing the proximal phalanx of the 2nd toe into this thumb.

DWL/SD

1.8.79

After discussion with mother I have advised that we should take the proximal phalanx from his 4th toe and inserted it to the thumb. This would then be fixed with a k wire. I have indicated that when this was done by Dr. Carroll it usually regenerates satisfactorily. Mother has agreed to this.

DWL/SD

2.8.79

OPERATION - Transfer of bone from left 4th toe proximal phalanx to right hand thumb.

Surgeon - Mr. D.W. Lamb Anaes - Dr. Park
Assts - Dr. R. Boome
Dr. K. Erzingatzian

Procedure -

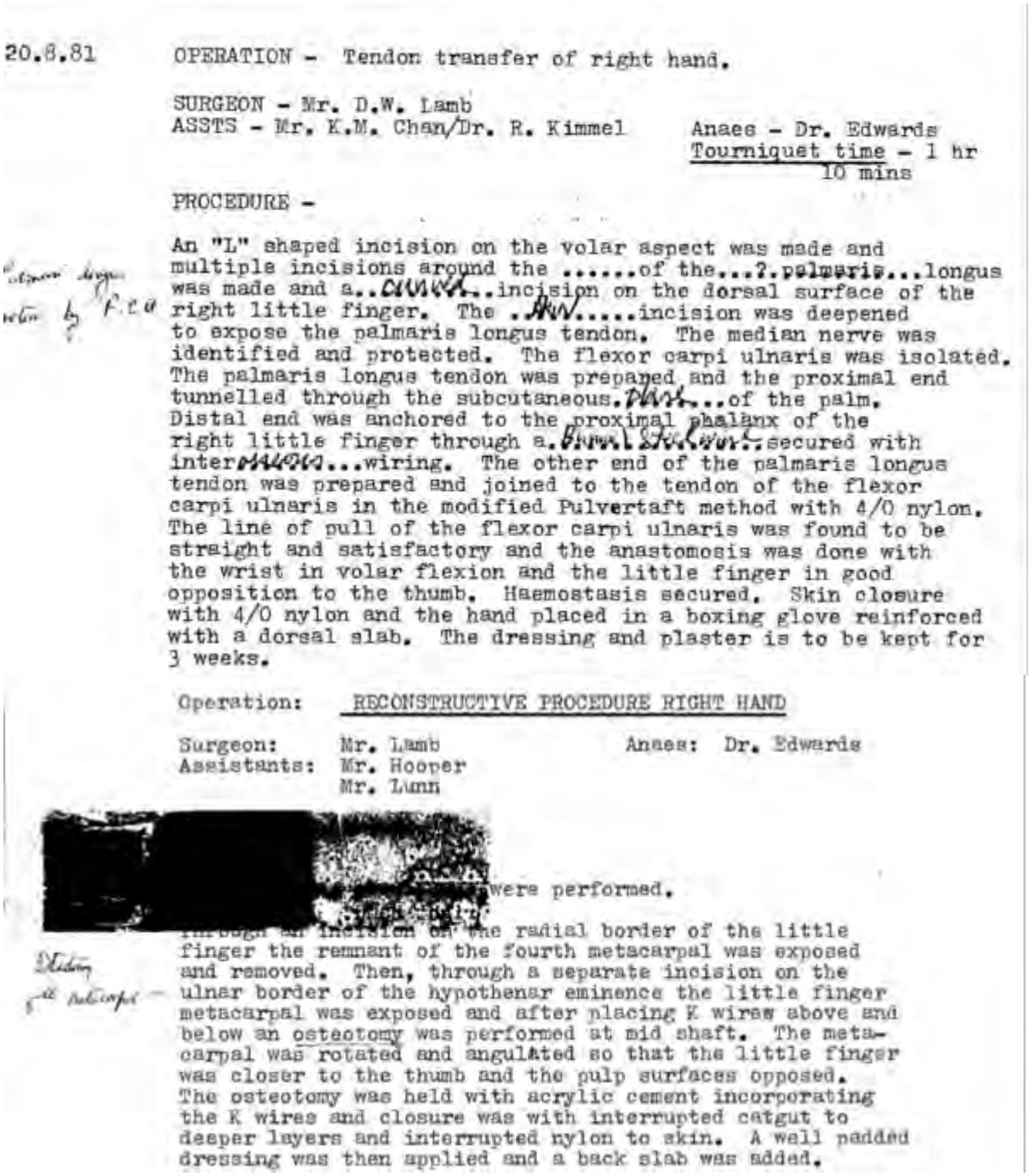
Dorsal zigzag incision on thumb. Potential space between distal phalanx and metacarpal exposed and extensor tendon cut. There was no flexor tendon, space gradually stretched. Soft tissue released until it was about ¾ cm in length. The distal end of the metacarpal was trimmed square. With removal of about 2-3 mm of cartilage the proximal end of the distal phalanx was trimmed square with removal of about 1 mm of cartilage. The proximal phalanx of the 4th toe on the left foot was removed subperiosteally through a dorsal oblique incision and periosteal .....closed with 3/0 chromic. Skin in interrupted nylon. Proximal phalanx was maintained intact and was inserted in the space between the metacarpal and distal phalanx of the right thumb with retrograde Kirschner wire extending from tip of thumb to metacarpal. There was a lot of tension in achieving this without trimming the graft bone at all and it was positioned with the angle at metacarpal graft to be about 30 deg. .....wards. The cartilage was not removed from the graft phalanx. It was not possible to reconstitute the extensor tendon to the thumb and the soft tissues were sutured around the graft, and the skin closed with interrupted nylon. A well padded plaster slab was applied. Before the skin was closed cuff was released and the finger pinked up well and haemostasis was obtained prior to closure.

Tourniquet time - 1 hr 10 mins for the hand.

RB/SD

Figure 51: Operation notes for the above patient with symbrachydactyly of the hand rather than the original diagnosis of 'cleft hand'.

CASE 30 (continued): *Symbrachydactyly (Hand Plate)*



20.8.81 OPERATION - Tendon transfer of right hand.

SURGEON - Mr. D.W. Lamb
ASSTS - Mr. K.M. Chan/Dr. R. Kimmel Anaes - Dr. Edwards
Tourniquet time - 1 hr 10 mins

PROCEDURE -

An "L" shaped incision on the volar aspect was made and multiple incisions around the .....of the...?.palmaris...longus was made and a..curved..incision on the dorsal surface of the right little finger. The ..this.....incision was deepened to expose the palmaris longus tendon. The median nerve was identified and protected. The flexor carpi ulnaris was isolated. The palmaris longus tendon was prepared and the proximal end tunnelled through the subcutaneous..plane...of the palm. Distal end was anchored to the proximal phalanx of the right little finger through a..burr hole.. secured with inter..mucos...wiring. The other end of the palmaris longus tendon was prepared and joined to the tendon of the flexor carpi ulnaris in the modified Pulvertaft method with 4/0 nylon. The line of pull of the flexor carpi ulnaris was found to be straight and satisfactory and the anastomosis was done with the wrist in volar flexion and the little finger in good opposition to the thumb. Haemostasis secured. Skin closure with 4/0 nylon and the hand placed in a boxing glove reinforced with a dorsal slab. The dressing and plaster is to be kept for 3 weeks.

Operation: RECONSTRUCTIVE PROCEDURE RIGHT HAND

Surgeon: Mr. Lamb Anaes: Dr. Edwards
Assistants: Mr. Hooper
Mr. Lunn

[redacted] were performed.

[redacted] through an incision on the radial border of the little finger the remnant of the fourth metacarpal was exposed and removed. Then, through a separate incision on the ulnar border of the hypothenar eminence the little finger metacarpal was exposed and after placing K wires above and below an osteotomy was performed at mid shaft. The metacarpal was rotated and angulated so that the little finger was closer to the thumb and the pulp surfaces opposed. The osteotomy was held with acrylic cement incorporating the K wires and closure was with interrupted catgut to deeper layers and interrupted nylon to skin. A well padded dressing was then applied and a back slab was added.

Figure 51 (continued): Operation notes for the above patient with symbrachydactyly of the hand rather than the original diagnosis of 'cleft hand'.

CASE 31: *Symbrachydactyly or Transverse Arrest (Hand Plate)*

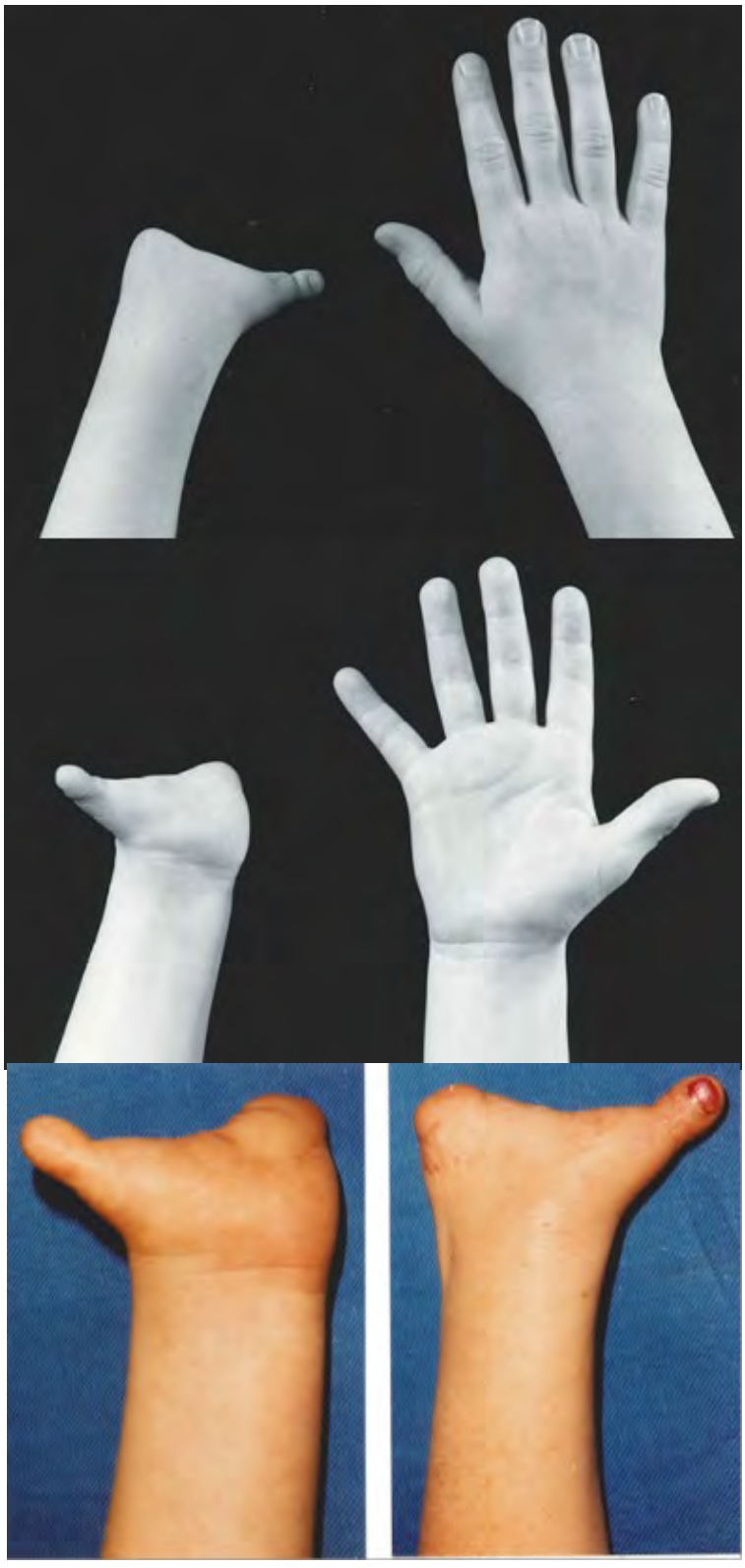
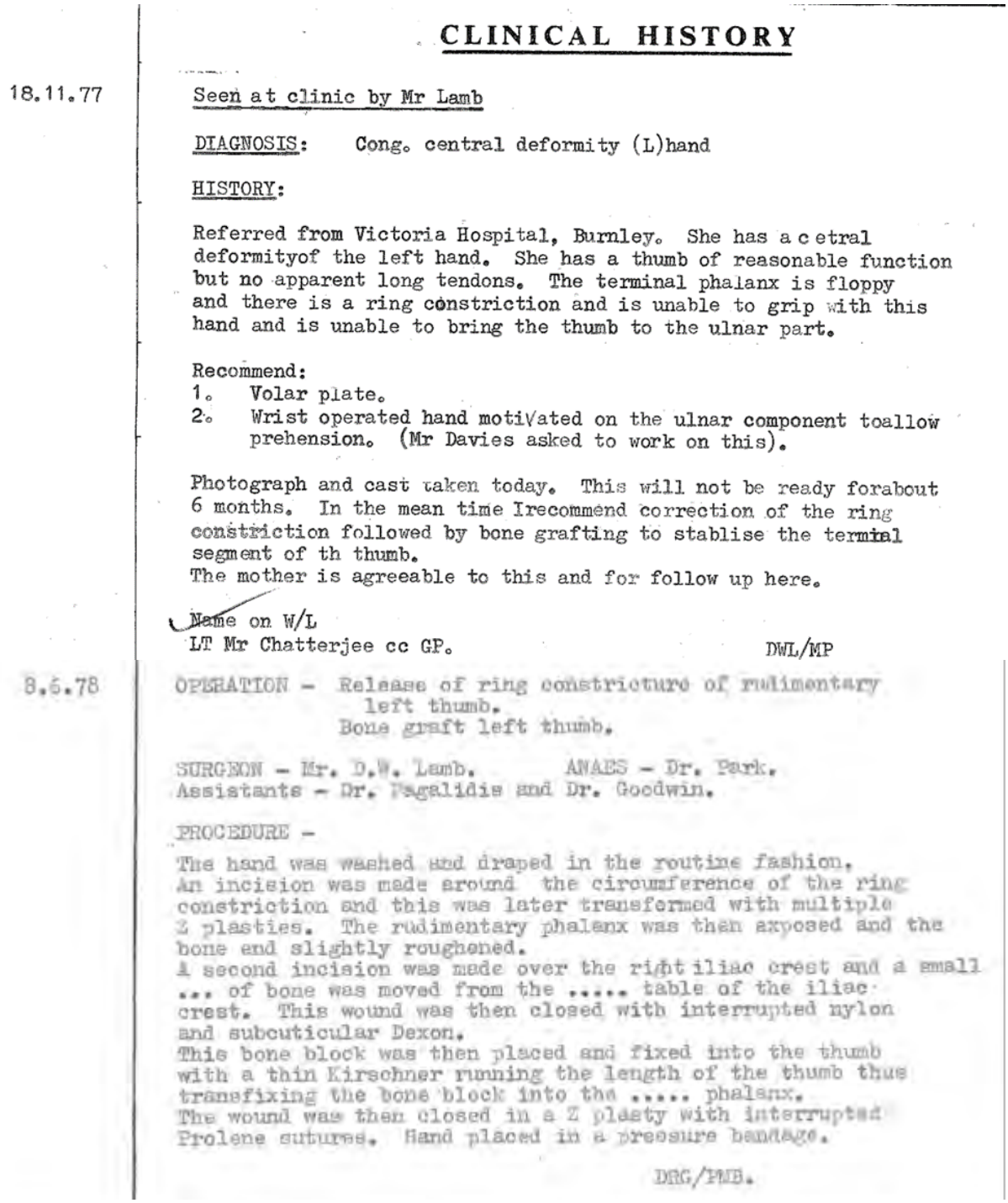


Figure 52: This patient had an original diagnosis of congenital central deformity left hand. The diagnosis is probably one of a proximal hand transverse arrest or a severe symbrachydactyly. Another diagnosis given was that of ring constriction syndrome.

This was probably derived from a ring around the rudimentary left thumb, although this could be a typical appearance of a hypoplastic thumb in symbrachydactyly.

CASE 31 (continued): *Symbrachydactyly or Transverse Arrest (Hand Plate)*



CLINICAL HISTORY

18.11.77

Seen at clinic by Mr Lamb

DIAGNOSIS: Cong. central deformity (L)hand

HISTORY:

Referred from Victoria Hospital, Burnley. She has a c etral deformityof the left hand. She has a thumb of reasonable function but no apparent long tendons. The terminal phalanx is floppy and there is a ring constriction and is unable to grip with this hand and is unable to bring the thumb to the ulnar part.

Recommend:
1. Volar plate.
2. Wrist operated hand motivated on the ulnar component toallow prehension. (Mr Davies asked to work on this).

Photograph and cast taken today. This will not be ready forabout 6 months. In the mean time Irecommend correction of the ring constriction followed by bone grafting to stablise the terminal segment of th thumb.
The mother is agreeable to this and for follow up here.

Name on W/L
LT Mr Chatterjee cc GP. DWL/MP

8.6.78

OPERATION - Release of ring constricture of rudimentary left thumb.
Bone graft left thumb.

SURGEON - Mr. D.W. Lamb. ANAES - Dr. Park.
Assistants - Dr. Pagalidis and Dr. Goodwin.

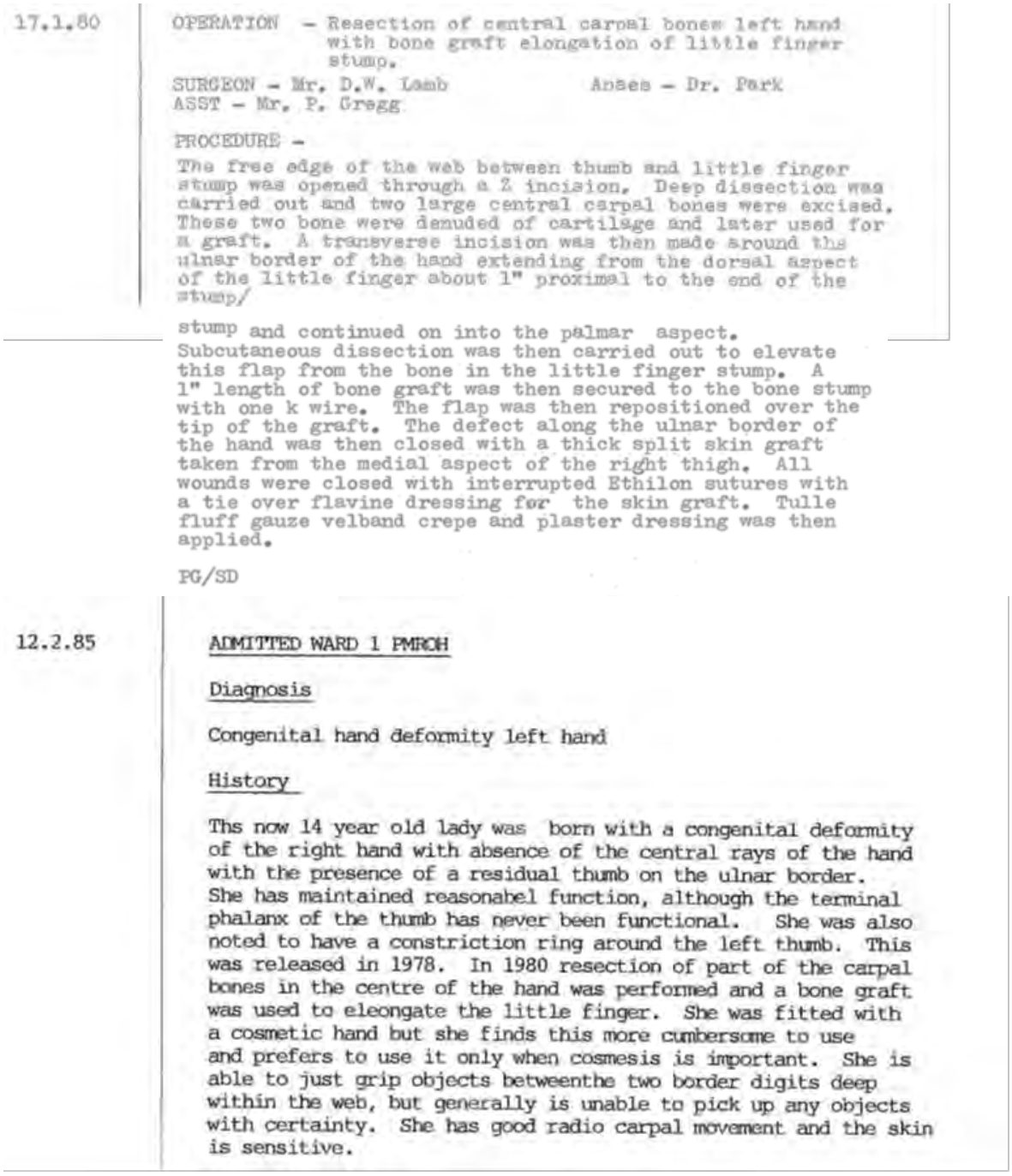
PROCEDURE -

The hand was washed and draped in the routine fashion.
An incision was made around the circumference of the ring constriction and this was later transformed with multiple Z plasties. The rudimentary phalanx was then exposed and the bone end slightly roughened.
A second incision was made over the right iliac crest and a small ... of bone was moved from the ..... table of the iliac crest. This wound was then closed with interrupted nylon and subcuticular Dexon.
This bone block was then placed and fixed into the thumb with a thin Kirschner running the length of the thumb thus transfixing the bone block into the ..... phalanx.
The wound was then closed in a Z plasty with interrupted Prolene sutures. Hand placed in a pressure bandage.

DRG/PMB.

Figure 53: This patient underwent several operations, including release of the ring constriction of the thumb, carpus rotation osteotomy of the radial digit, and radial advancement of the ulnar digit.

CASE 31 (continued): *Symbrachydactyly or Transverse Arrest (Hand Plate)*



17.1.80

OPERATION – Resection of central carpal bones left hand with bone graft elongation of little finger stump.

SURGEON – Mr. D.W. Lamb Anaes – Dr. Park
ASST – Mr. P. Gregg

PROCEDURE –

The free edge of the web between thumb and little finger stump was opened through a Z incision. Deep dissection was carried out and two large central carpal bones were excised. These two bone were denuded of cartilage and later used for a graft. A transverse incision was then made around the ulnar border of the hand extending from the dorsal aspect of the little finger about 1" proximal to the end of the stump/

stump and continued on into the palmar aspect. Subcutaneous dissection was then carried out to elevate this flap from the bone in the little finger stump. A 1" length of bone graft was then secured to the bone stump with one k wire. The flap was then repositioned over the tip of the graft. The defect along the ulnar border of the hand was then closed with a thick split skin graft taken from the medial aspect of the right thigh. All wounds were closed with interrupted Ethilon sutures with a tie over flavine dressing for the skin graft. Tulle fluff gauze velband crepe and plaster dressing was then applied.

PG/SD

12.2.85

ADMITTED WARD 1 PMRCH

Diagnosis

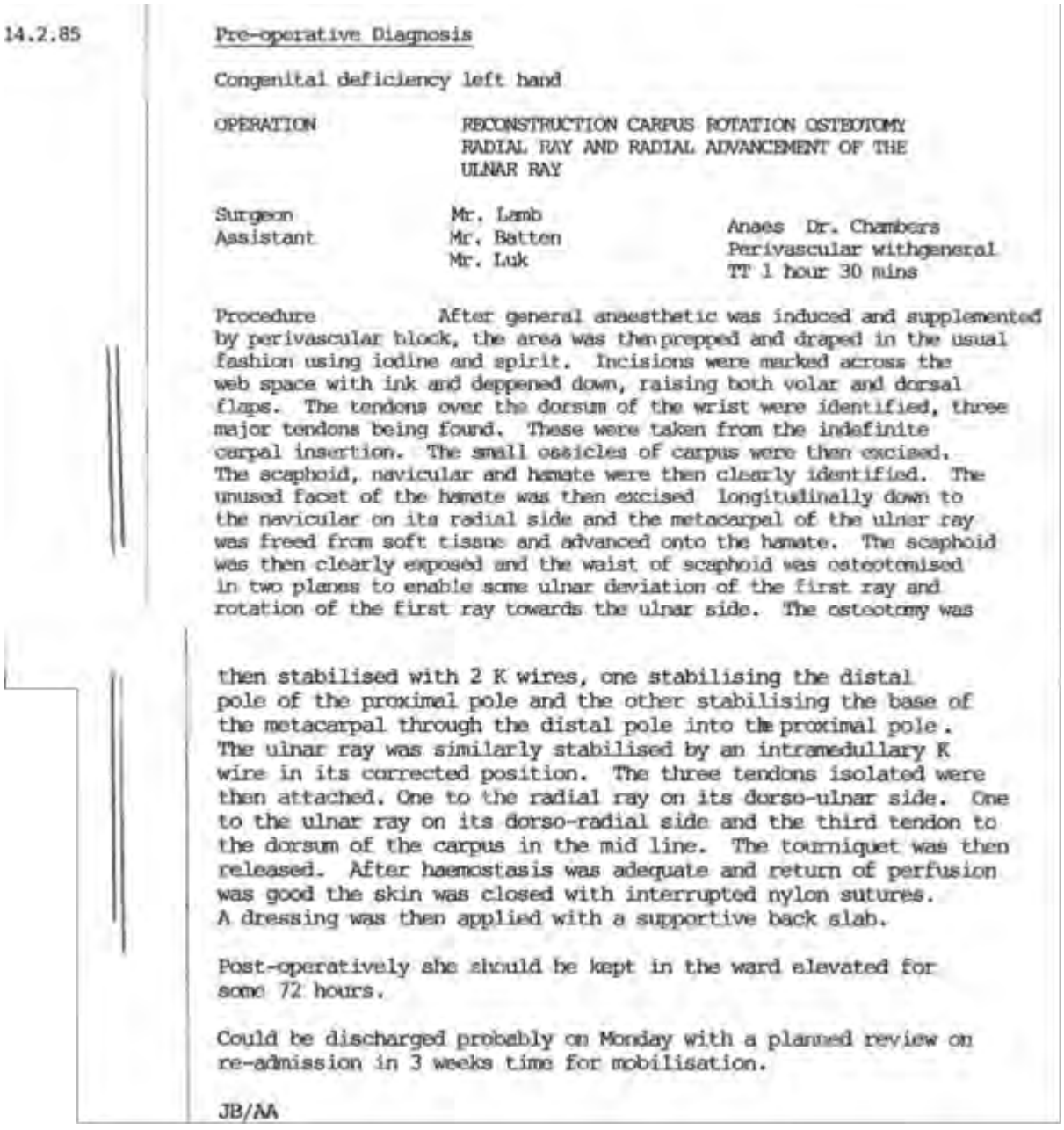
Congenital hand deformity left hand

History

Ths now 14 year old lady was born with a congenital deformity of the right hand with absence of the central rays of the hand with the presence of a residual thumb on the ulnar border. She has maintained reasonabel function, although the terminal phalanx of the thumb has never been functional. She was also noted to have a constriction ring around the left thumb. This was released in 1978. In 1980 resection of part of the carpal bones in the centre of the hand was performed and a bone graft was used to eleongate the little finger. She was fitted with a cosmetic hand but she finds this more cumbersome to use and prefers to use it only when cosmesis is important. She is able to just grip objects betweenthe two border digits deep within the web, but generally is unable to pick up any objects with certainty. She has good radio carpal movement and the skin is sensitive.

Figure 53 (continued): This patient underwent several operations, including release of the ring constriction of the thumb and also carpus rotation osteotomy of the radial digit and radial advancement of the ulnar digit.

CASE 31 (continued): *Symbrachydactyly or Transverse Arrest (Hand Plate)*



14.2.85

Pre-operative Diagnosis

Congenital deficiency left hand

OPERATION — RECONSTRUCTION CARPUS ROTATION OSTEOTOMY RADIAL RAY AND RADIAL ADVANCEMENT OF THE ULNAR RAY

Surgeon — Mr. Lamb
Assistant — Mr. Batten, Mr. Luk
Anaes Dr. Chambers
Perivascular withgeneral
TT 1 hour 30 mins

Procedure — After general anaesthetic was induced and supplemented by perivascular block, the area was then prepped and draped in the usual fashion using iodine and spirit. Incisions were marked across the web space with ink and deppened down, raising both volar and dorsal flaps. The tendons over the dorsum of the wrist were identified, three major tendons being found. These were taken from the indefinite carpal insertion. The small ossicles of carpus were then excised. The scaphoid, navicular and hamate were then clearly identified. The unused facet of the hamate was then excised longitudinally down to the navicular on its radial side and the metacarpal of the ulnar ray was freed from soft tissue and advanced onto the hamate. The scaphoid was then clearly exposed and the waist of scaphoid was osteotomised in two planes to enable some ulnar deviation of the first ray and rotation of the first ray towards the ulnar side. The osteotomy was

then stabilised with 2 K wires, one stabilising the distal pole of the proximal pole and the other stabilising the base of the metacarpal through the distal pole into the proximal pole. The ulnar ray was similarly stabilised by an intramedullary K wire in its corrected position. The three tendons isolated were then attached. One to the radial ray on its dorso-ulnar side. One to the ulnar ray on its dorso-radial side and the third tendon to the dorsum of the carpus in the mid line. The tourniquet was then released. After haemostasis was adequate and return of perfusion was good the skin was closed with interrupted nylon sutures. A dressing was then applied with a supportive back slab.

Post-operatively she should be kept in the ward elevated for some 72 hours.

Could be discharged probably on Monday with a planned review on re-admission in 3 weeks time for mobilisation.

JB/AA

Figure 53 (continued): This patient underwent several operations, including release of the ring constriction of the thumb, carpus rotation osteotomy of the radial digit, and radial advancement of the ulnar digit.

CASE 32: *Transverse Arrest (Hand Plate)*

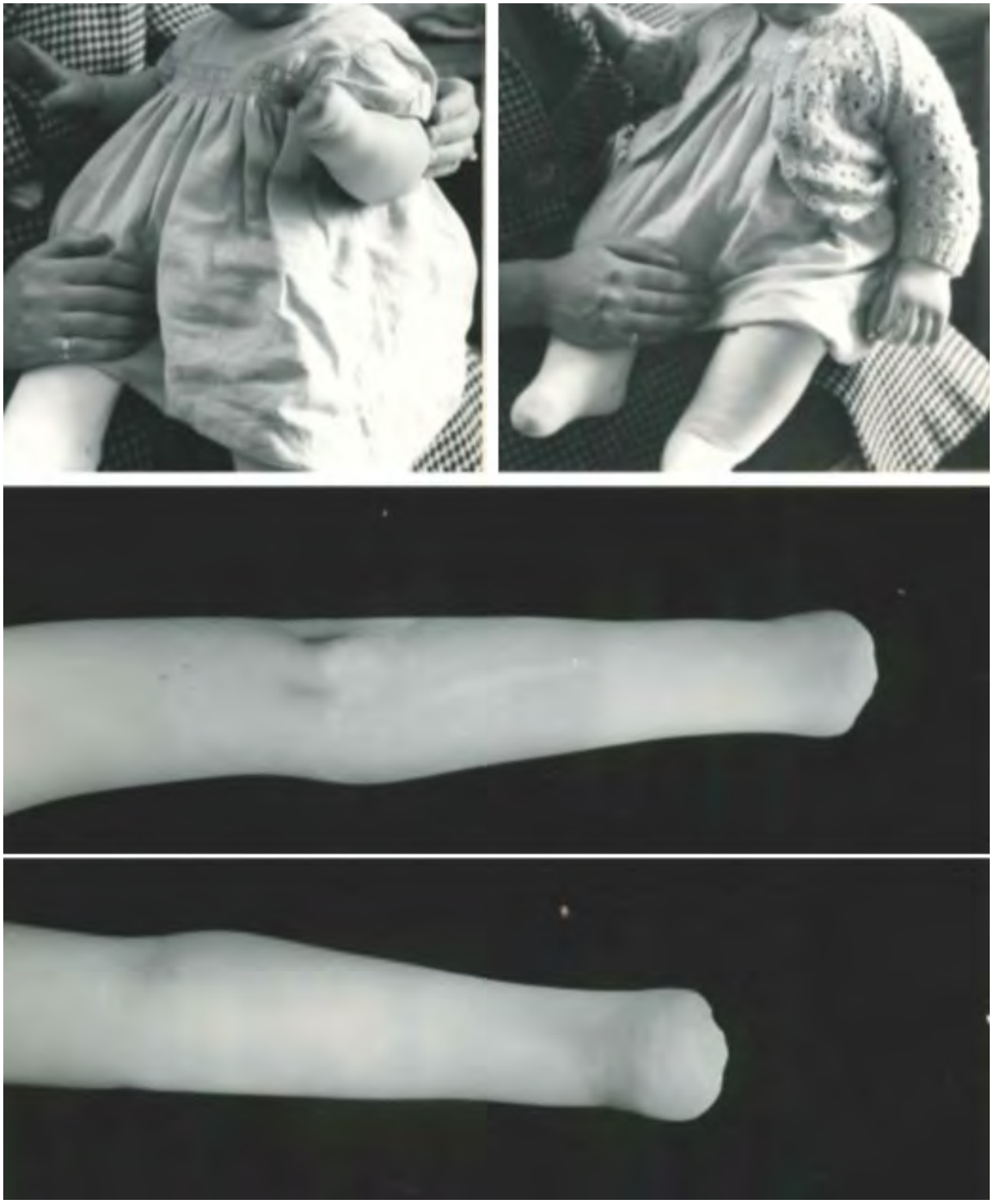


Figure 54: In this patient you can see a wrist joint, and after that very little growth of the hand with no evidence of nubbin. This is a transverse arrest of the proximal hand, and in her case she was fitted with a plastic one-piece limb prosthesis.

CASE 33: *Transverse Arrest (Hand Plate)*

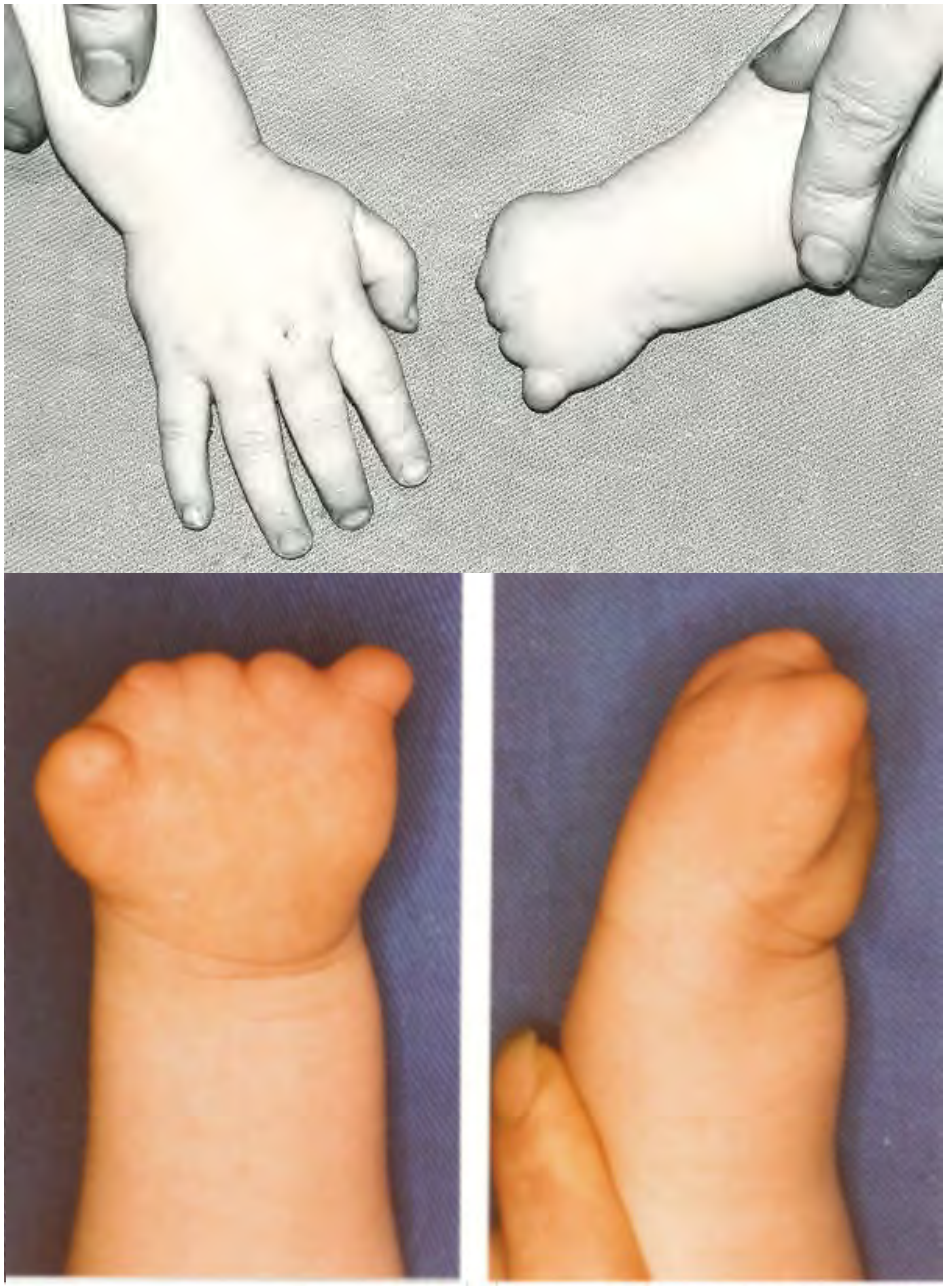


Figure 55: This is another case of left transverse arrest at a more distal level. Again the wrist joint is present and the fingers stopped growing around the metacarpophalangeal joint level. In this case, the patient underwent surgery for deepening of the first web space and excision of the second metacarpal in order to give her some grip prehension.

I-B-2-iv. Cleft Hand

THE CLEFT HAND is an interesting entity. The proper name is perhaps split hand and foot syndrome, of which there are at least seven types. The pathogenesis is believed to be a defect in the apical ectodermal ridge, preventing the growth of central structures as the hand is developing. Manske's classification is usually used, which focuses on the first web space. This is a useful classification which can guide treatment. In these children, their function is usually excellent, although the appearances can be quite upsetting. Adrian Flatt calls this "a functional triumph but a social disaster."

The term "lobster claw hand" was used several times in the notes to describe the cleft hand. This is a term that should not be used nowadays, but it was widely used for several decades.

CASE 34: *Cleft Hand*

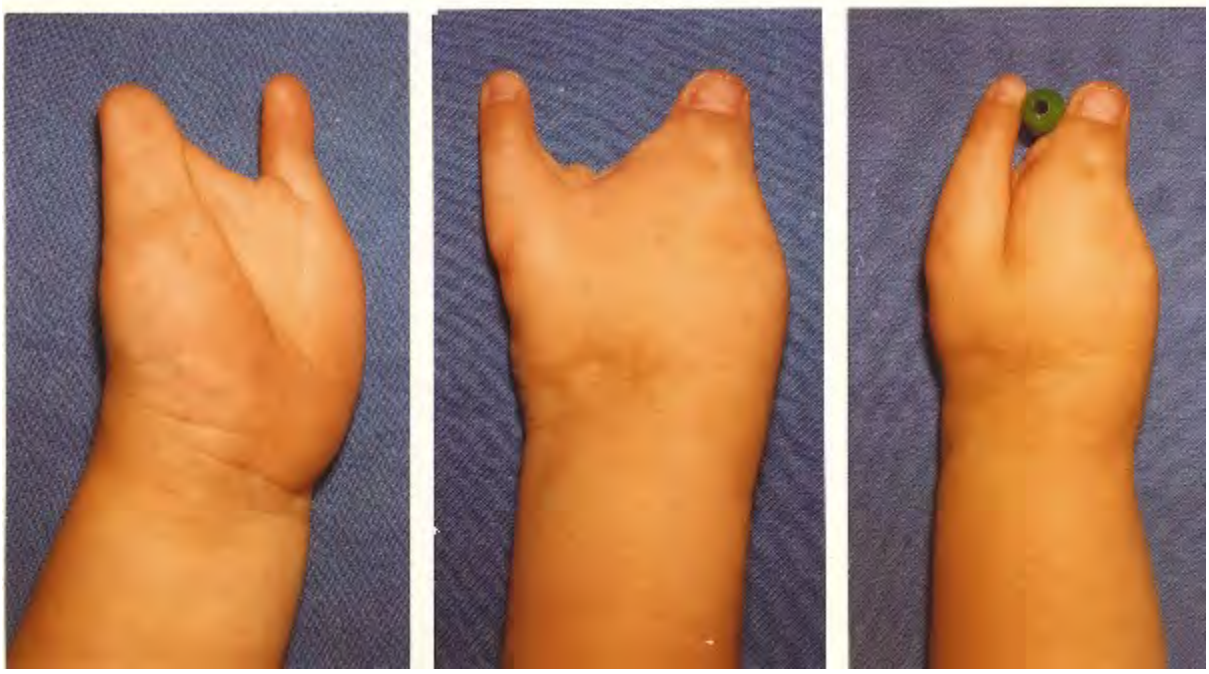


Figure 56: In this patient, the diagnosis of "lobster claw hand" was given. This is a severity cleft hand deformity where the radial digits have fused with the thumb leaving a single digit on the ulnar aspect. It is easy to see how some may debate the diagnosis of this condition to be symbrachydactyly or even ulnar dysplasia. In this case, instead of closing the cleft, Mr Lamb interestingly deepened the cleft and also performed a rotational osteotomy of the digits to gain a better prehension. He combined this with a sublimis opposition transfer to the left thumb as well.

CASE 34 (continued): *Cleft Hand*

PRINCESS MARGARET ROSE ORTHOPAEDIC HOSPITAL

NEGATIVE No.

MEDICAL PHOTOGRAPHIC DEPARTMENT

PATIENT AGE 7yrs WARD 1

DATE SURGEON MR LAMB

CLINICAL DIAGNOSIS LOBSTER CLAW HAND

THIS COPY IS FOR THE POCKET OF THE PATIENT'S NOTES

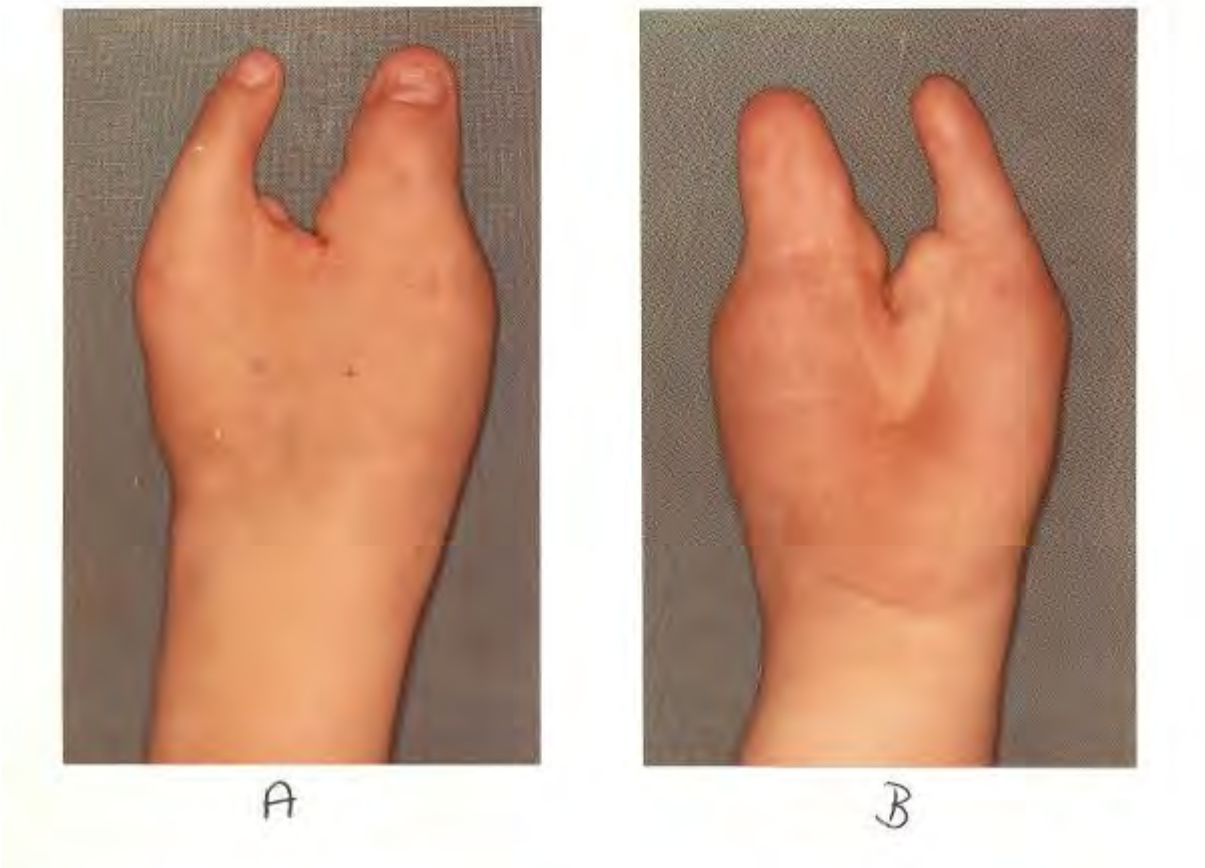


Figure 57: Post-op photographs of the above patient with cleft hand.

CASE 35: *Cleft Hand*

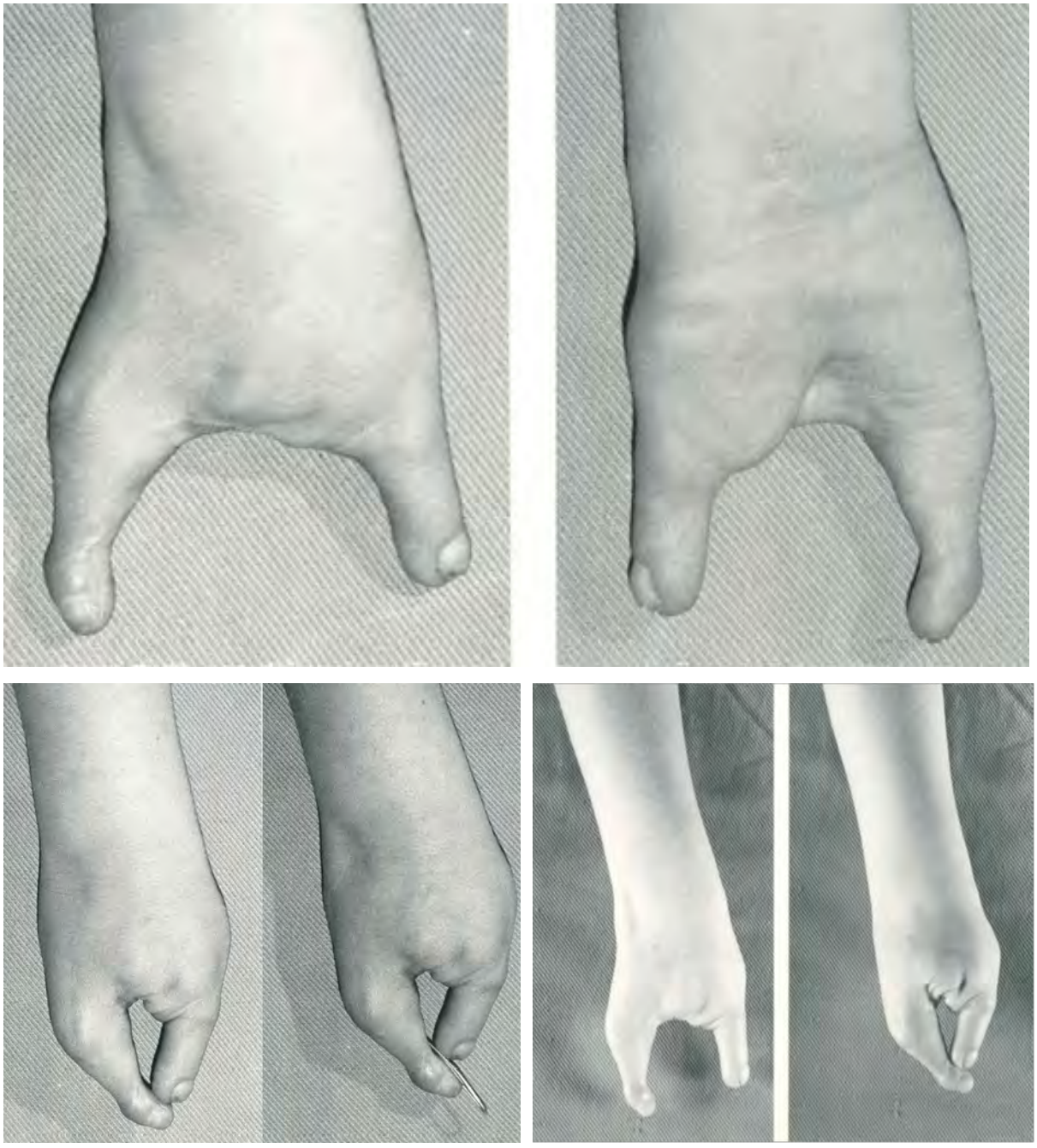
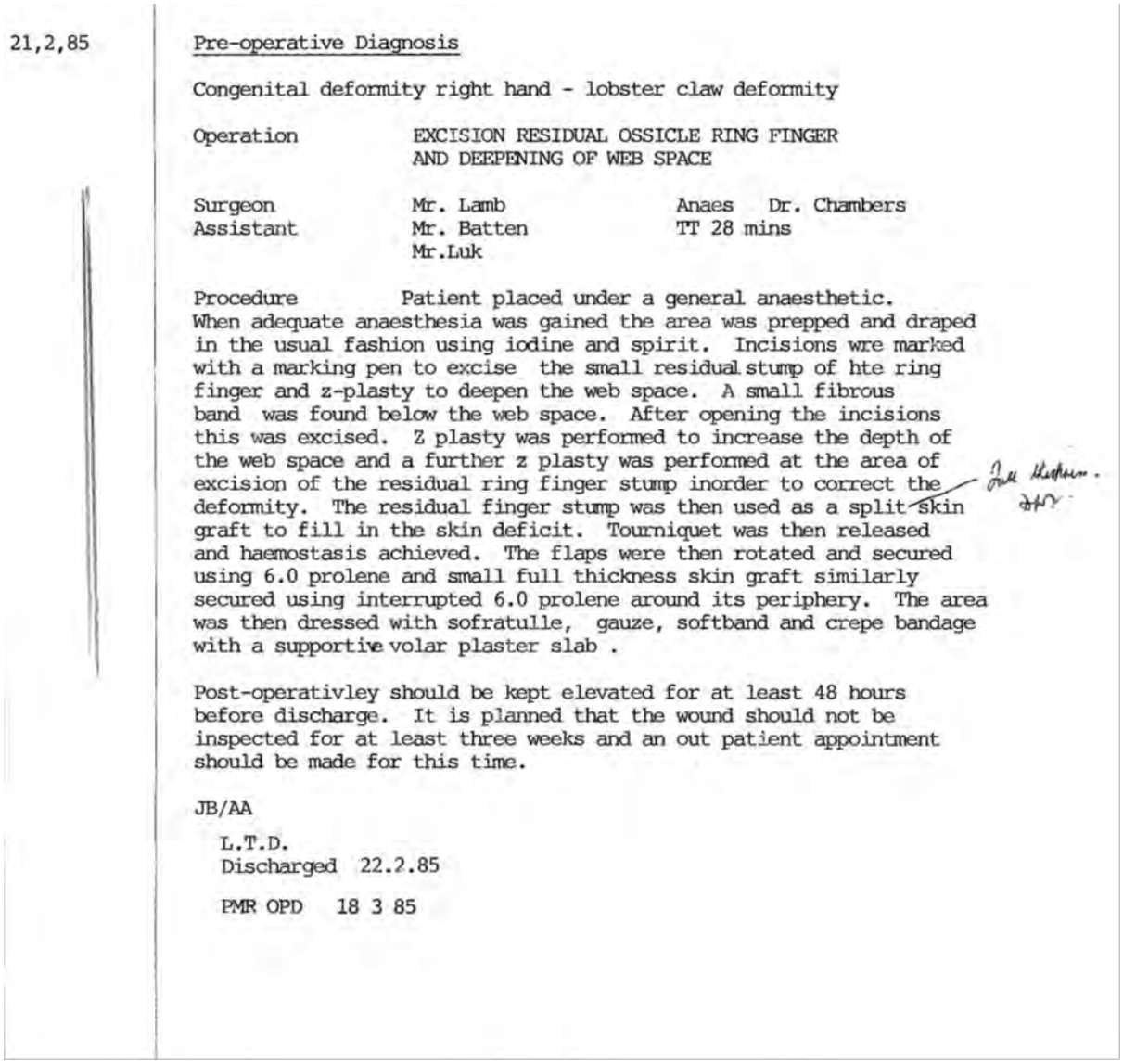


Figure 58: This is another case that was described as a "lobster claw hand" on the right side. She was described as having excellent function. Therefore, no surgical treatment was indicated. She also had a pectoral deficiency consistent with Poland syndrome. Subsequently she had a nubbin excised, which may suggest that the diagnosis could also be symbrachydactyly.

CASE 35 (continued): *Cleft Hand*



21,2,85

Pre-operative Diagnosis

Congenital deformity right hand - lobster claw deformity

Operation EXCISION RESIDUAL OSSICLE RING FINGER
AND DEEPENING OF WEB SPACE

Surgeon Mr. Lamb Anaes Dr. Chambers
Assistant Mr. Batten TT 28 mins
Mr.Luk

Procedure Patient placed under a general anaesthetic. When adequate anaesthesia was gained the area was prepped and draped in the usual fashion using iodine and spirit. Incisions wre marked with a marking pen to excise the small residual stump of hte ring finger and z-plasty to deepen the web space. A small fibrous band was found below the web space. After opening the incisions this was excised. Z plasty was performed to increase the depth of the web space and a further z plasty was performed at the area of excision of the residual ring finger stump inorder to correct the deformity. The residual finger stump was then used as a split skin graft to fill in the skin deficit. Tourniquet was then released and haemostasis achieved. The flaps were then rotated and secured using 6.0 prolene and small full thickness skin graft similarly secured using interrupted 6.0 prolene around its periphery. The area was then dressed with sofratulle, gauze, softband and crepe bandage with a supportive volar plaster slab .

Post-operativley should be kept elevated for at least 48 hours before discharge. It is planned that the wound should not be inspected for at least three weeks and an out patient appointment should be made for this time.

JB/AA

L.T.D.
Discharged 22.2.85

PMR OPD 18 3 85

Figure 59: Operation notes for the above patient with cleft hand who underwent an excision of a nubbin.

I-B-2. Malformation of the Hand Plate:

Radioulnar Axis

SIMILAR TO the malformations of the entire upper limb, defects in the radioulnar axis of the hand plate are secondary to various mutations affecting the zone of polarising activity or its downstream activities. The Sonic hedgehog protein pathway is the central mechanism for differentiations of tissues in the radioulnar axis as well as digit patterning.

In this category, conditions such as thumb hypoplasia, polydactyly affecting both the preaxial and postaxial axes are predominant.

Hypoplastic thumb (radial longitudinal deficiency). As mentioned, certain conditions seldom affect the hand in isolation. In this condition it is important to examine the hypoplastic thumb in conjunction with the forearm. Frequently even in the absence of an obvious radial deviation, the forearm is shortened. The associated syndromes of radial longitudinal deficiency must also be borne in mind, ie VACTERL or Fanconi anaemia. Hypoplastic thumb is popularly classified by the Blauth classification. More accurately, this is the Manske's modification of Blauth's classification, and it is used to describe different levels of severity of thumb hypoplasia.

CASE 36: *Hypoplastic Thumb*

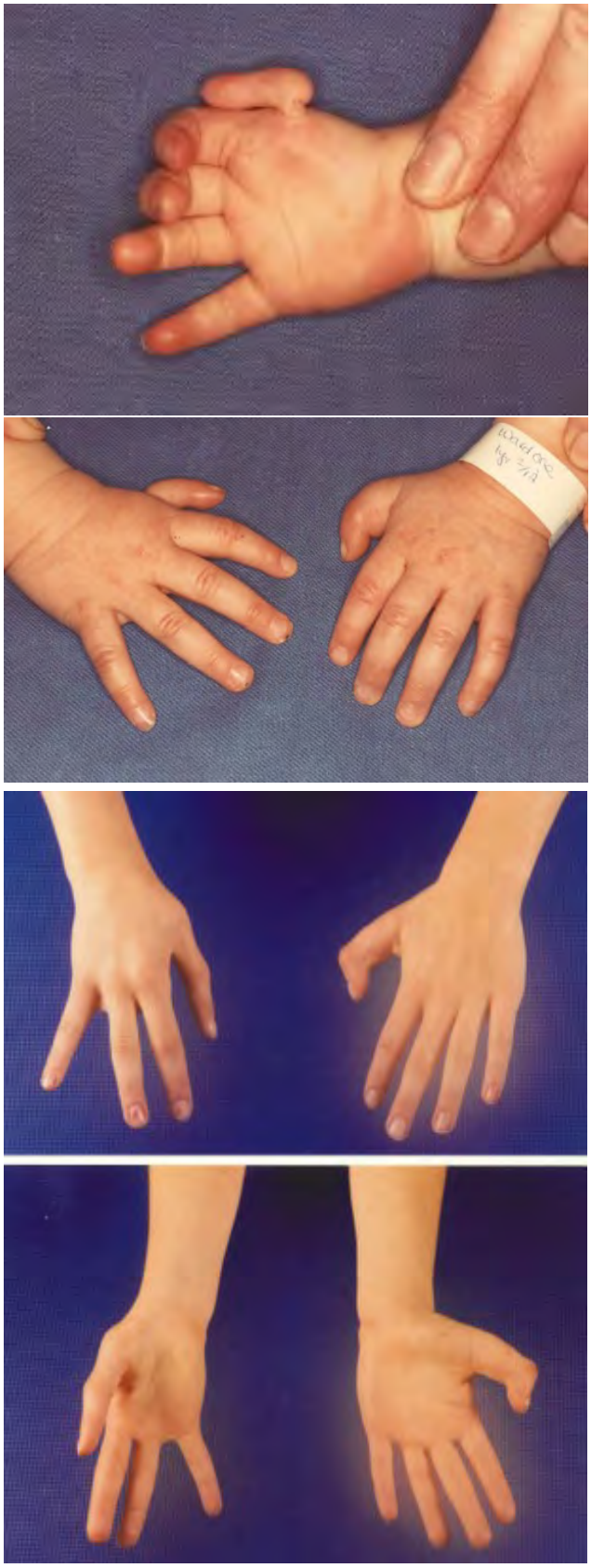


Figure 60: This patient has a right Blauth 4 thumb hypoplasia, which is a floating thumb. He underwent resection of the radial duplicate digit and an index finger pollicisation. The way Douglas Lamb did his pollicisation has been described earlier. It is interesting to study the operation notes to see the specifics of his technique. He seemed to prefer the use of chromic catgut sutures rather than K-wires when rotating the index finger into the desired position.

CASE 37: *Hypoplastic Thumb*

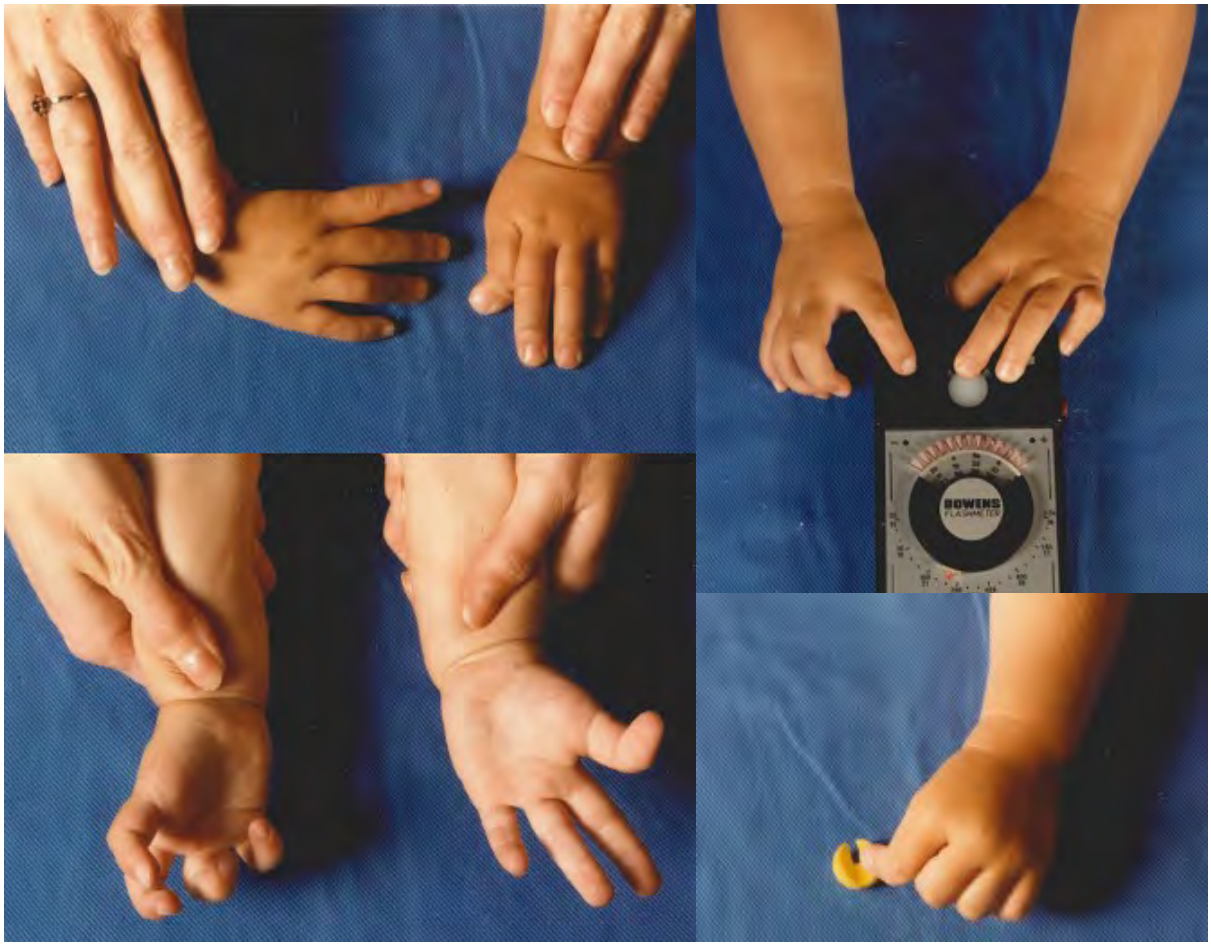
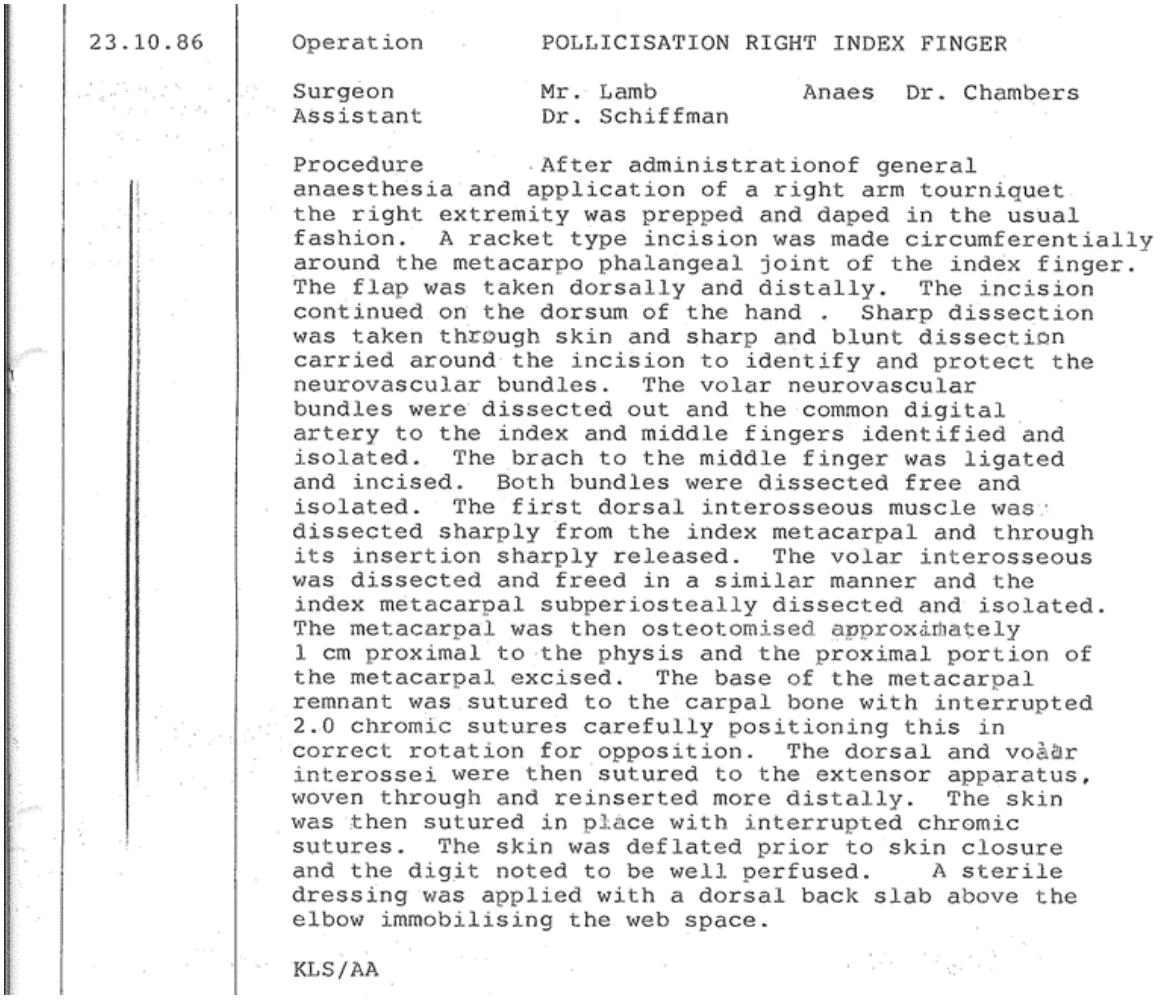


Figure 61: This is a patient with bilateral thumb hypoplasia. In both hands, these are Blauth type 5 deformities. He underwent a pollicisation on the right index finger. On the left he had correction of the deviation followed by a pollicisation of the left index finger.

CASE 37 (continued): *Hypoplastic Thumb*



23.10.86

Operation POLLICISATION RIGHT INDEX FINGER

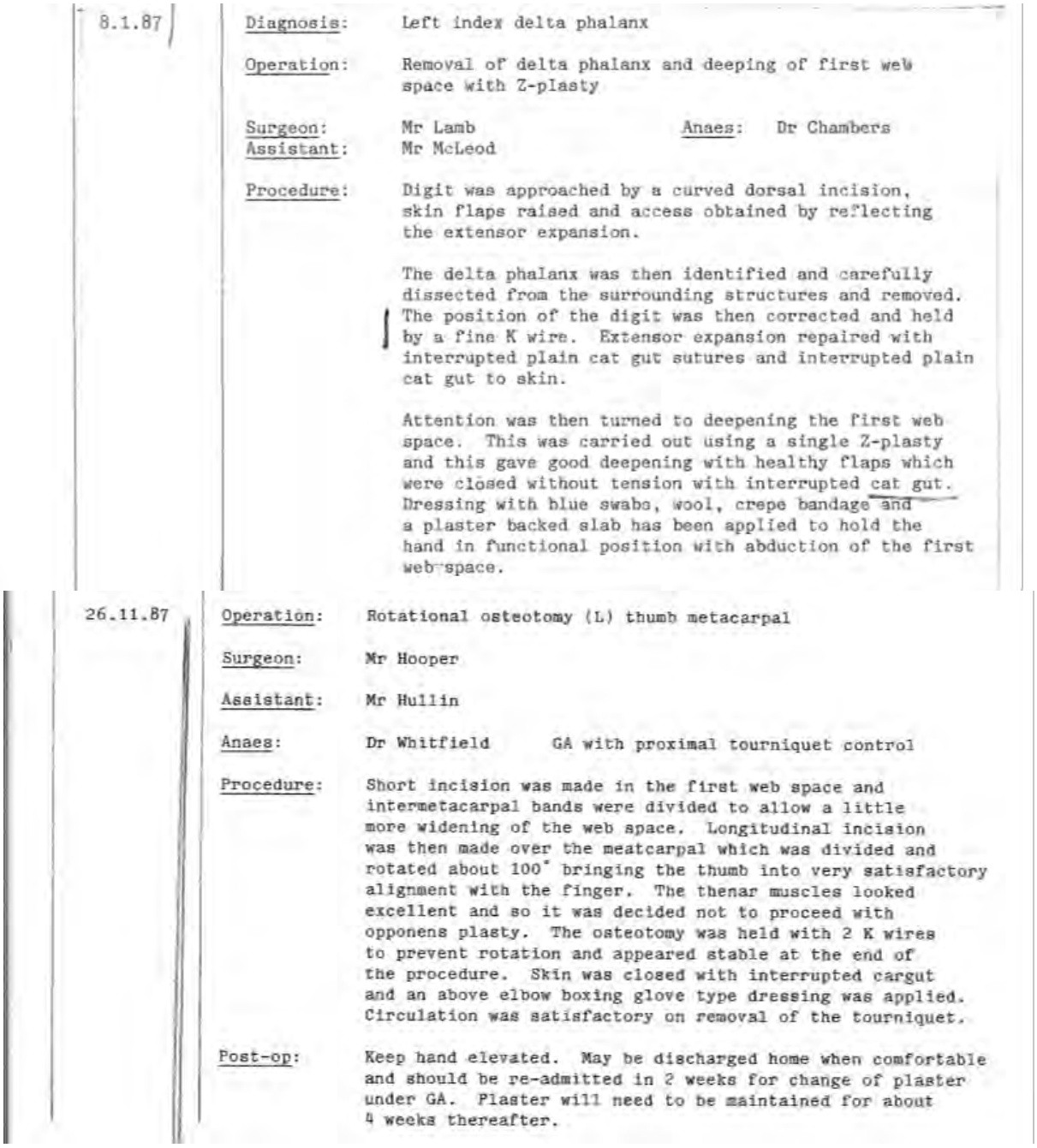
Surgeon Mr. Lamb Anaes Dr. Chambers
Assistant Dr. Schiffman

Procedure After administrationof general anaesthesia and application of a right arm tourniquet the right extremity was prepped and daped in the usual fashion. A racket type incision was made circumferentially around the metacarpo phalangeal joint of the index finger. The flap was taken dorsally and distally. The incision continued on the dorsum of the hand . Sharp dissection was taken through skin and sharp and blunt dissection carried around the incision to identify and protect the neurovascular bundles. The volar neurovascular bundles were dissected out and the common digital artery to the index and middle fingers identified and isolated. The brach to the middle finger was ligated and incised. Both bundles were dissected free and isolated. The first dorsal interosseous muscle was dissected sharply from the index metacarpal and through its insertion sharply released. The volar interosseous was dissected and freed in a similar manner and the index metacarpal subperiosteally dissected and isolated. The metacarpal was then osteotomised approximately 1 cm proximal to the physis and the proximal portion of the metacarpal excised. The base of the metacarpal remnant was sutured to the carpal bone with interrupted 2.0 chromic sutures carefully positioning this in correct rotation for opposition. The dorsal and voàar interossei were then sutured to the extensor apparatus, woven through and reinserted more distally. The skin was then sutured in place with interrupted chromic sutures. The skin was deflated prior to skin closure and the digit noted to be well perfused. A sterile dressing was applied with a dorsal back slab above the elbow immobilising the web space.

KLS/AA

Figure 62: Operation notes for the above patient with bilateral thumb hypoplasia.

CASE 37 (continued): *Hypoplastic Thumb*



8.1.87

Diagnosis: Left index delta phalanx

Operation: Removal of delta phalanx and deeping of first web space with Z-plasty

Surgeon: Mr Lamb Anaes: Dr Chambers
Assistant: Mr McLeod

Procedure: Digit was approached by a curved dorsal incision, skin flaps raised and access obtained by reflecting the extensor expansion.

The delta phalanx was then identified and carefully dissected from the surrounding structures and removed. The position of the digit was then corrected and held by a fine K wire. Extensor expansion repaired with interrupted plain cat gut sutures and interrupted plain cat gut to skin.

Attention was then turned to deepening the first web space. This was carried out using a single Z-plasty and this gave good deepening with healthy flaps which were closed without tension with interrupted cat gut. Dressing with blue swabs, wool, crepe bandage and a plaster backed slab has been applied to hold the hand in functional position with abduction of the first web space.

26.11.87

Operation: Rotational osteotomy (L) thumb metacarpal

Surgeon: Mr Hooper

Assistant: Mr Hullin

Anaes: Dr Whitfield GA with proximal tourniquet control

Procedure: Short incision was made in the first web space and intermetacarpal bands were divided to allow a little more widening of the web space. Longitudinal incision was then made over the meatcarpal which was divided and rotated about 100° bringing the thumb into very satisfactory alignment with the finger. The thenar muscles looked excellent and so it was decided not to proceed with opponens plasty. The osteotomy was held with 2 K wires to prevent rotation and appeared stable at the end of the procedure. Skin was closed with interrupted cargut and an above elbow boxing glove type dressing was applied. Circulation was satisfactory on removal of the tourniquet.

Post-op: Keep hand elevated. May be discharged home when comfortable and should be re-admitted in 2 weeks for change of plaster under GA. Plaster will need to be maintained for about 4 weeks thereafter.

Figure 62 (continued): Operation notes for the above patient with bilateral thumb hypoplasia.

CASE 38: *Hypoplastic Thumb*

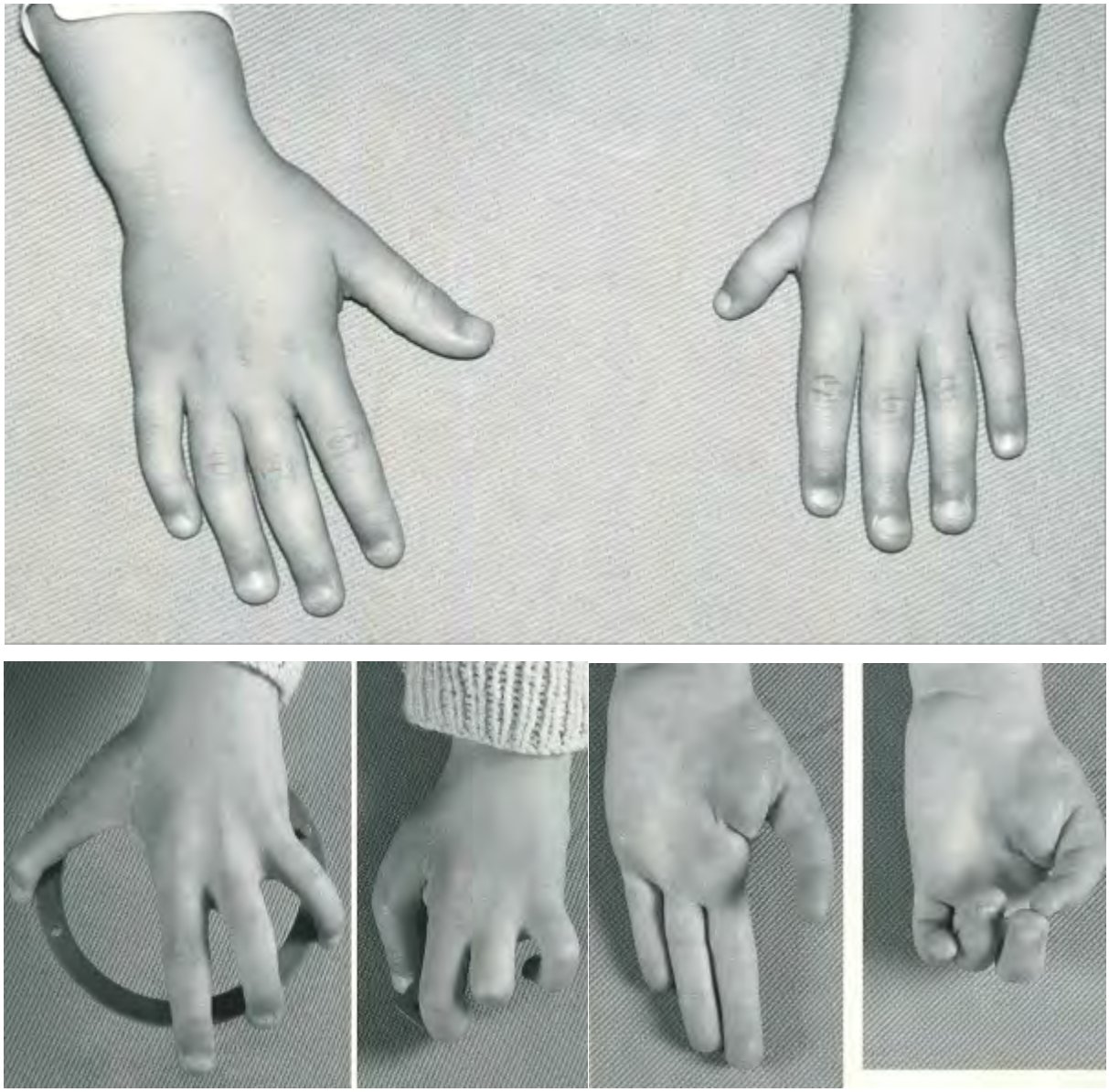


Figure 63: This patient has what is likely to be a Blauth 3B thumb on the left hand, and he underwent a pollicisation.

CASE 39: *Hypoplastic Thumb*

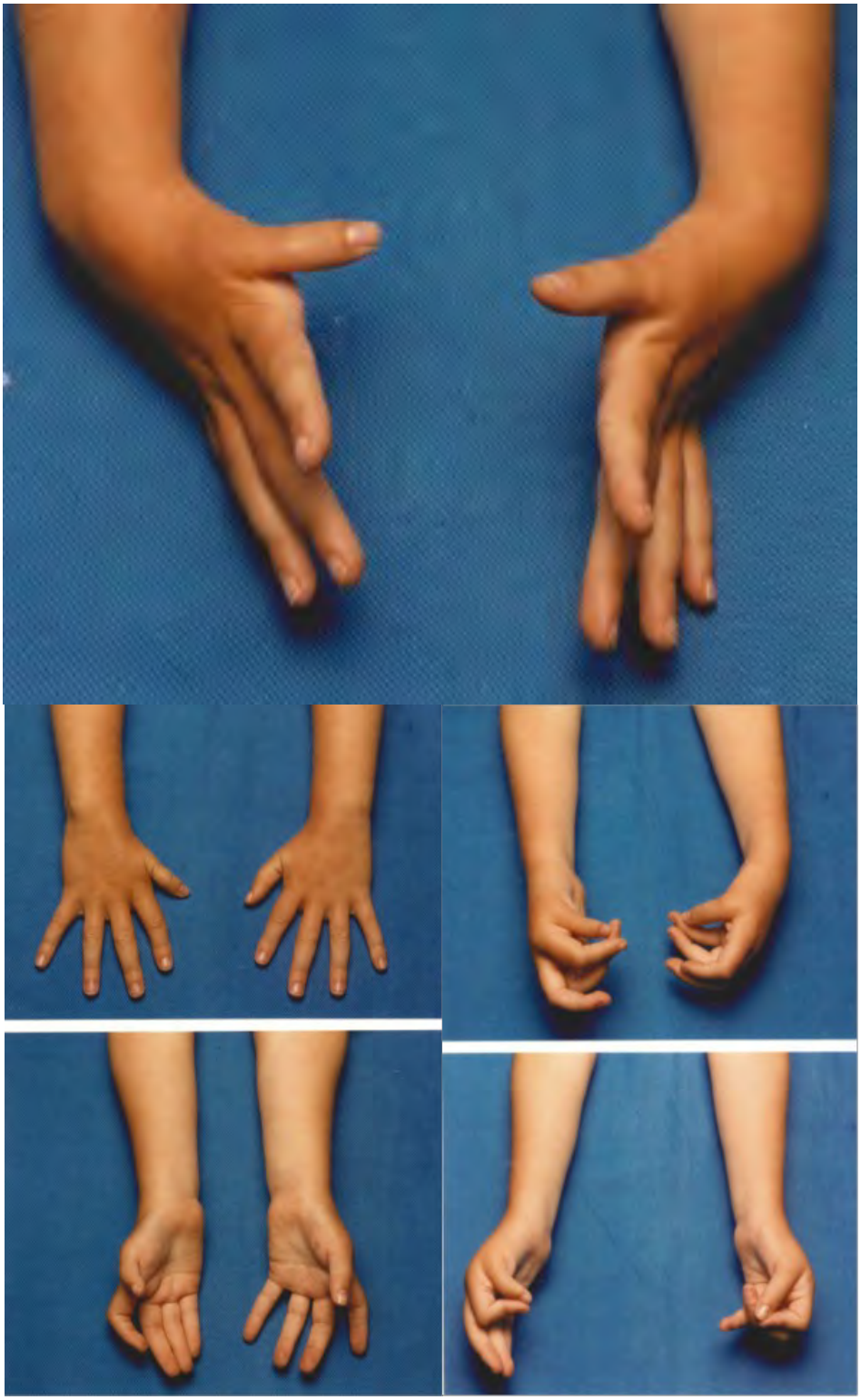


Figure 64: This is probably a left thumb hypoplasia. Patient underwent a tendon transfer for improved opposition.

CASE 40: *Thumb Duplication*

2.9.76

OPERATION : Amputation accessory radial left thumb.

SURGEON : Mr. D.W. Lamb. ASSISTANTS : Dr. S. Smith
ANAES : Dr. Park. Mr. J. Cochrane

PROCEDURE :

Zig zag incision over dorsal ~~of~~ volar aspects radial accessory digit left thumb. The additional phalanx was dissected out and removed. There was no bony synostosis.

The major EPL and FPL tendons both went to the ulnar thumb. A radial strip of the extensor pollicis longus tendon was mobilised from distally tp proximally and this together with the small remnant of EPL which passed to the accessory radial thumb, was passed beneath extensor pollicis longus and transferred into the radial side of this tendon to try and prevent the interphalangeal joint of the thumb from drifting into the position of radial deviation.

Wound closed with interrupted catgut to deep fascia, with interrupted catgut to fat and with interrupted nylon sutures to skin.
½Sofra tulle dressing, vellband, crepe bandage and POP back shell applied.

Figure 65: This patient has a left thumb duplication, which was removed. It was difficult to know which was the classification. The thumb was described as 'bifid'. The radial portion of finger has single phalanx, and ulnar two phalanges. It was also described as, "The thumb is tending to fall into radial division at the IP joint." The photograph depicted the thumb following removal of the duplicate but before wedge osteotomy to correct the radial deviation.

CASE 40 (continued): *Thumb Duplication*

29.3.79

OPERATION: Wedge osteotomy proximal phalanx left thumb with correction of deformities MP and IP joints and k wire fixation.

Surgeon: Mr. D.W. Lamb Assee: Dr. Park

PROCEDURE:

With G.A. and tourniquet control the previous scar on the dorso-radial aspect of the proximal phalanx left thumb was reopened and extended distally to mid-distal phalanx. The extensor apparatus was reflected from the radial aspect of the MP joint the proximal phalanx and IP joint and the attitude of the articular aspect of the head of the proximal phalanx shown to be some 40 deg. radial angulation. A closing wedge osteotomy in the middle of the proximal phalanx was therefore fashioned based on its ulnar aspect and with its closure the deviation was corrected. K wire was first passed proximally across the osteotomy site distally back through the distal phalanx then proximally again to stabilise the MP joint with correction of its ulnar deviation.

The extensor apparatus was replaced with plain catgut sutures and the skin closed with interrupted nylon. K-wire cut short and the wound dressed with Sofratulle and padded woollen crepe dressing complimented by plaster slab.

IC/SD

Postop. x-rays to be taken in Recovery Room. Note should be made that excess cartilage was removed from the distal radial aspect of the proximal phalanx during the operation.

When the Postoperative swelling has settled a plaster bridge could be fashioned across the unprotected end of the k wire then the patient permitted home. K wire to remain in situ for 6 weeks at least.

IC/SD

30.3.79

A satisfactory straightening out of the thumb was achieved. The articular surface at the distal inter-phalangeal joint was grossly distorted and could only be straightened into a reasonable position by wedge osteotomy through the distal phalanx. K wire fixation is necessary.

Patient is comfortable, circulation good. Check x-ray satisfactory. Can probably be allowed home tomorrow.

DWL/SD

Figure 66: There is a move towards immediate osteotomy to achieve a straighter thumb and to reduce the incidence of revision surgery following thumb duplication surgery. Despite this, there is always a risk for revision surgery due to the deforming forces in thumb duplication disorders.

CASE 41: *Triphalangeal Thumb*

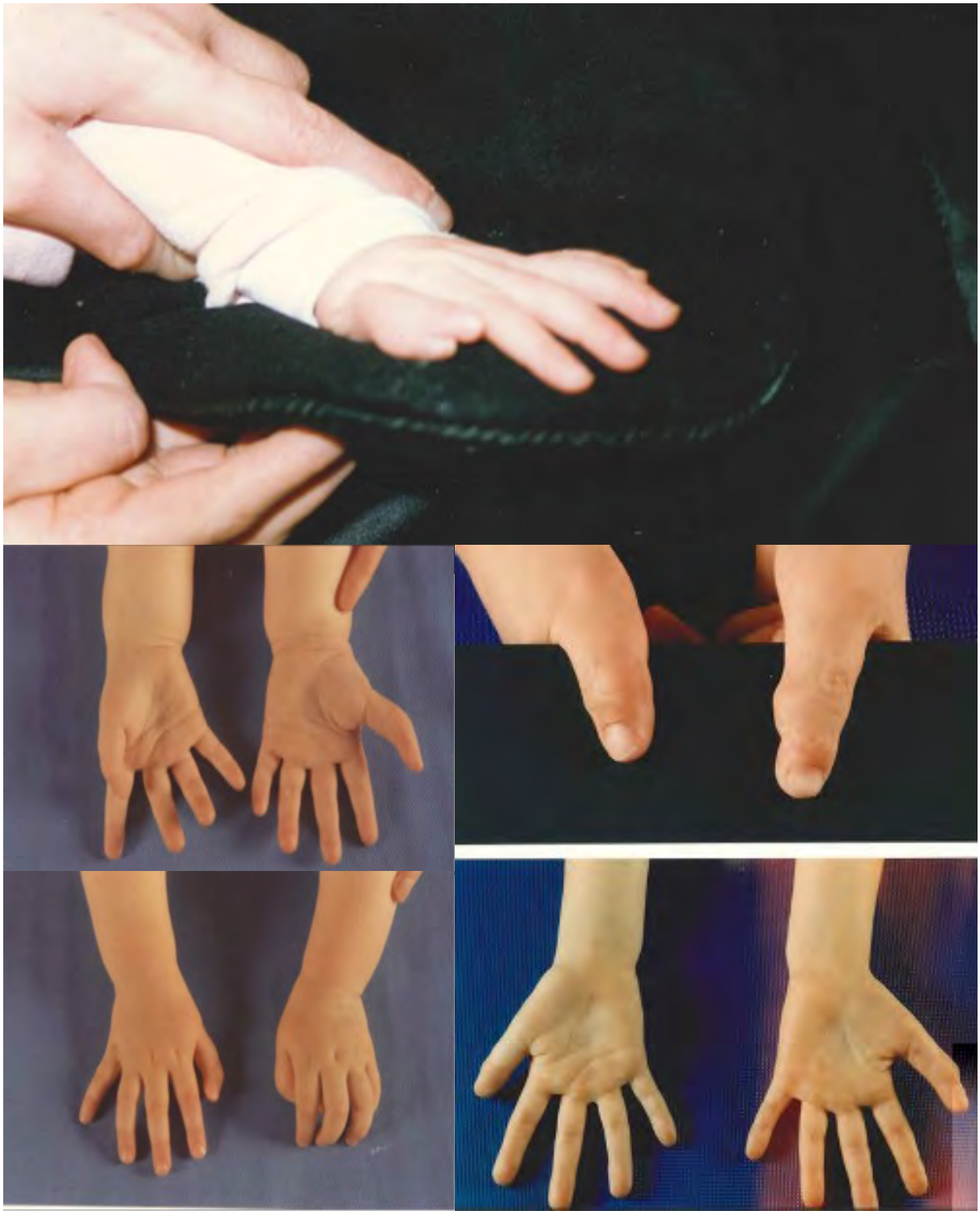


Figure 67: This is a case of triphalangeal thumb with radial duplication on the left. She underwent removal of the radial duplication on the left hand, and on the right removal of the delta phalanx to convert this from a triphalangeal thumb to a biphalangeal thumb.

CASE 42: *Five-finger Hand*

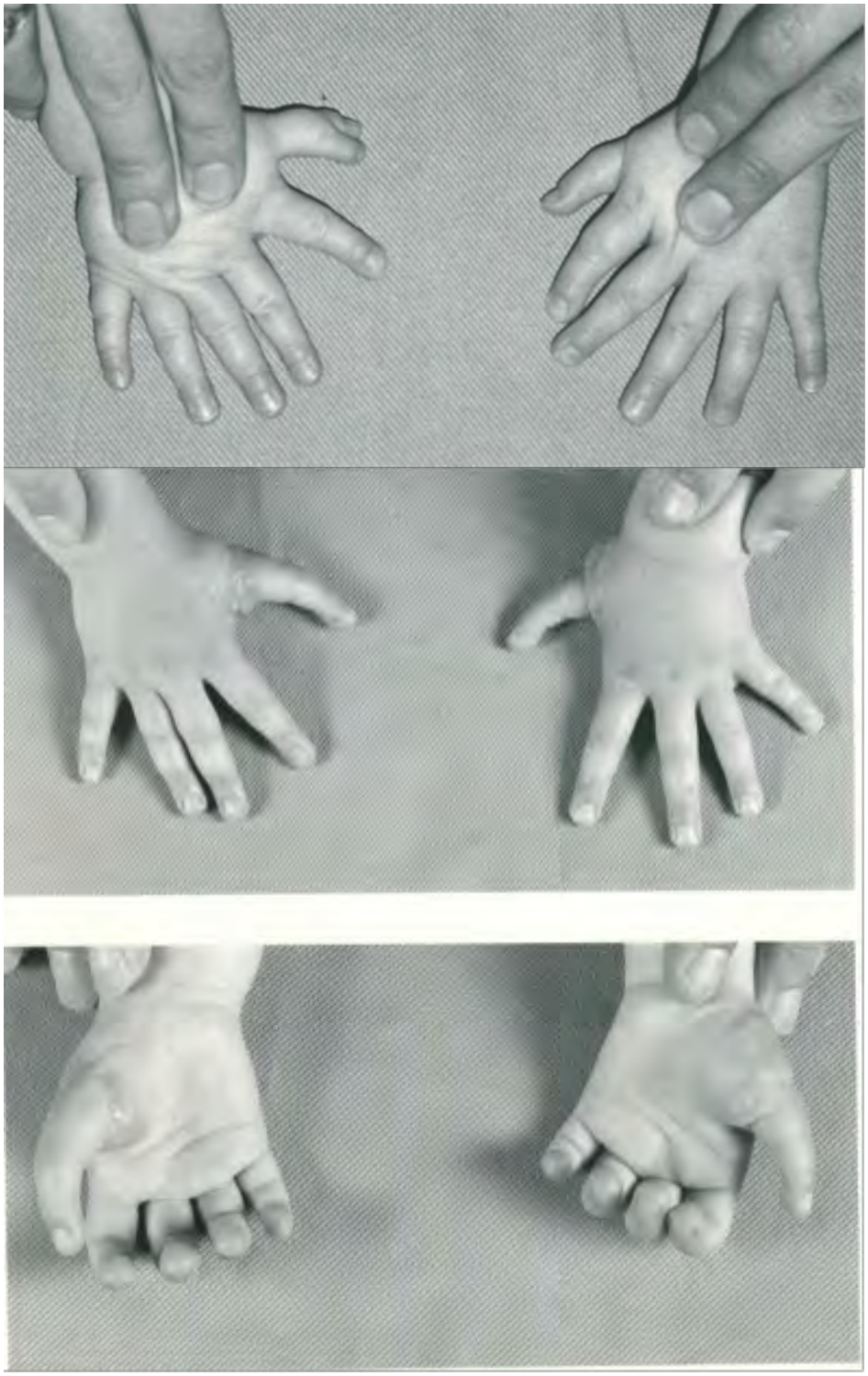


Figure 68: This is a patient with bilateral five-finger hands and various configurations of radial polydactyly. He underwent removal of extra digit on the right hand and pollicisation to obtain better hand function.

CASE 43: *Five-finger Hand*

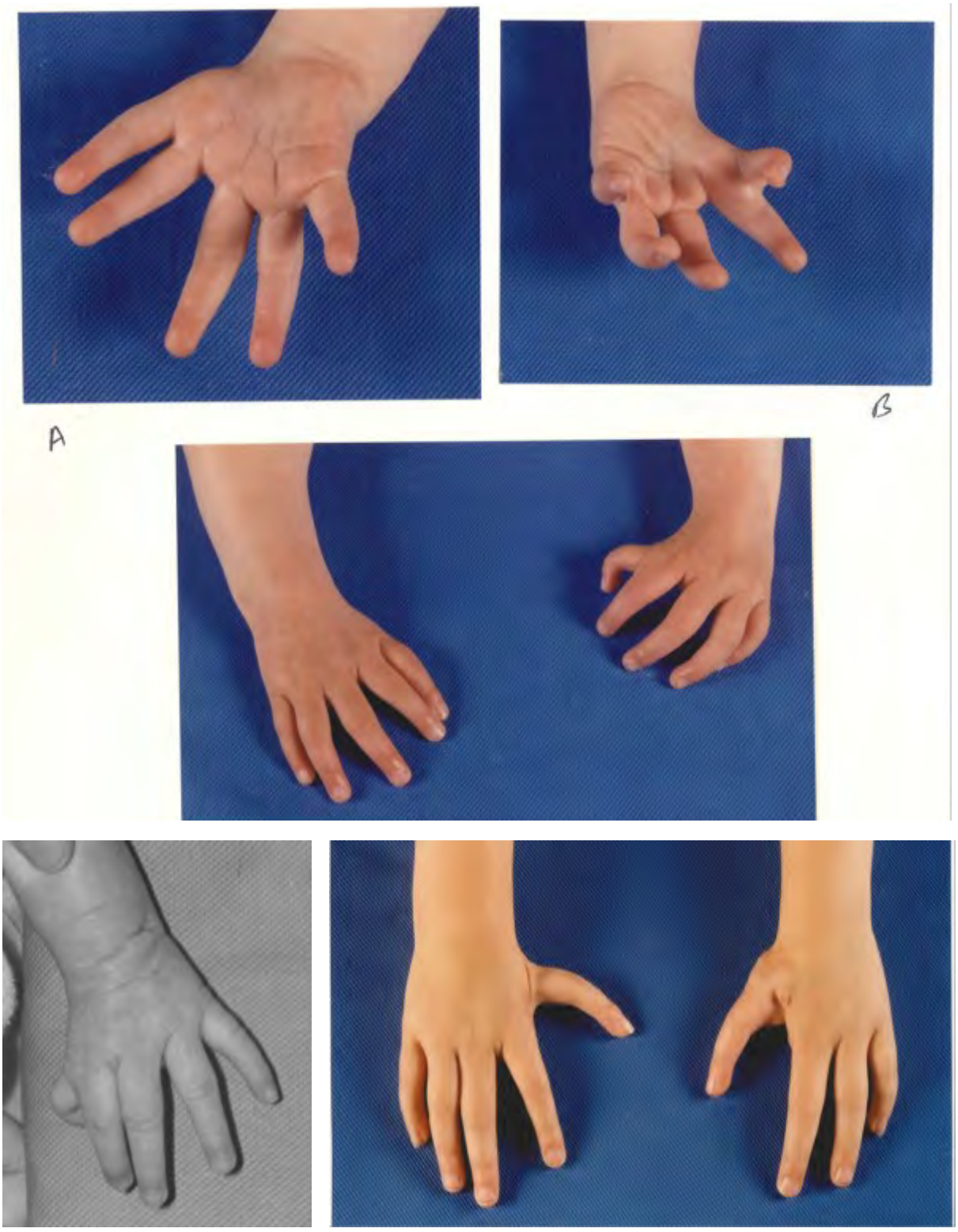


Figure 69: This patient underwent pollicisation of the radial digit on the right, followed by amputation of the radial digit on the left and pollicisation of the index finger

CASE 44: *Clinodactyly*

CLINICAL HISTORY

6.2.87

SEEN AT CLINIC BY MR LAMB

DIAGNOSIS :

HISTORY :

Referred by Mr Bissett because of culry little fignerm.
There is a familial tendency on maternal side.

There is a short middle phalanx with varus. I do not think it is likley to interfere with function.

No treatment meantime.
LT Mr Bissett cc GP.
DWL/MW

Figure 70: This is the one case of clinodactyly that we found in the notes. It was described as "curly little fingers." This patient was not treated surgically. The management of this condition has not changed much over the decades. There may be a move towards an earlier procedure to break up the epiphysis with a piece of fat, otherwise known as the Vickers procedure.

II. DEFORMATIONS

II-A. Deformations: ***Constriction Ring Sequence***

THIS IS the second major category of the OMT classification. Deformations refer to abnormal forces acting on a limb after it has been formed. The major group of conditions in this category are limbs or digits affected by constriction bends. There is a variety of terminologies for this condition, including amniotic band sequence or constriction ring sequence if [mild] digits or limbs present with a circular ring of indentations with no distal lymphedema. If severe, this can lead to auto-amputation of limbs/digits.

CASE 45: *Constriction Ring Sequence*

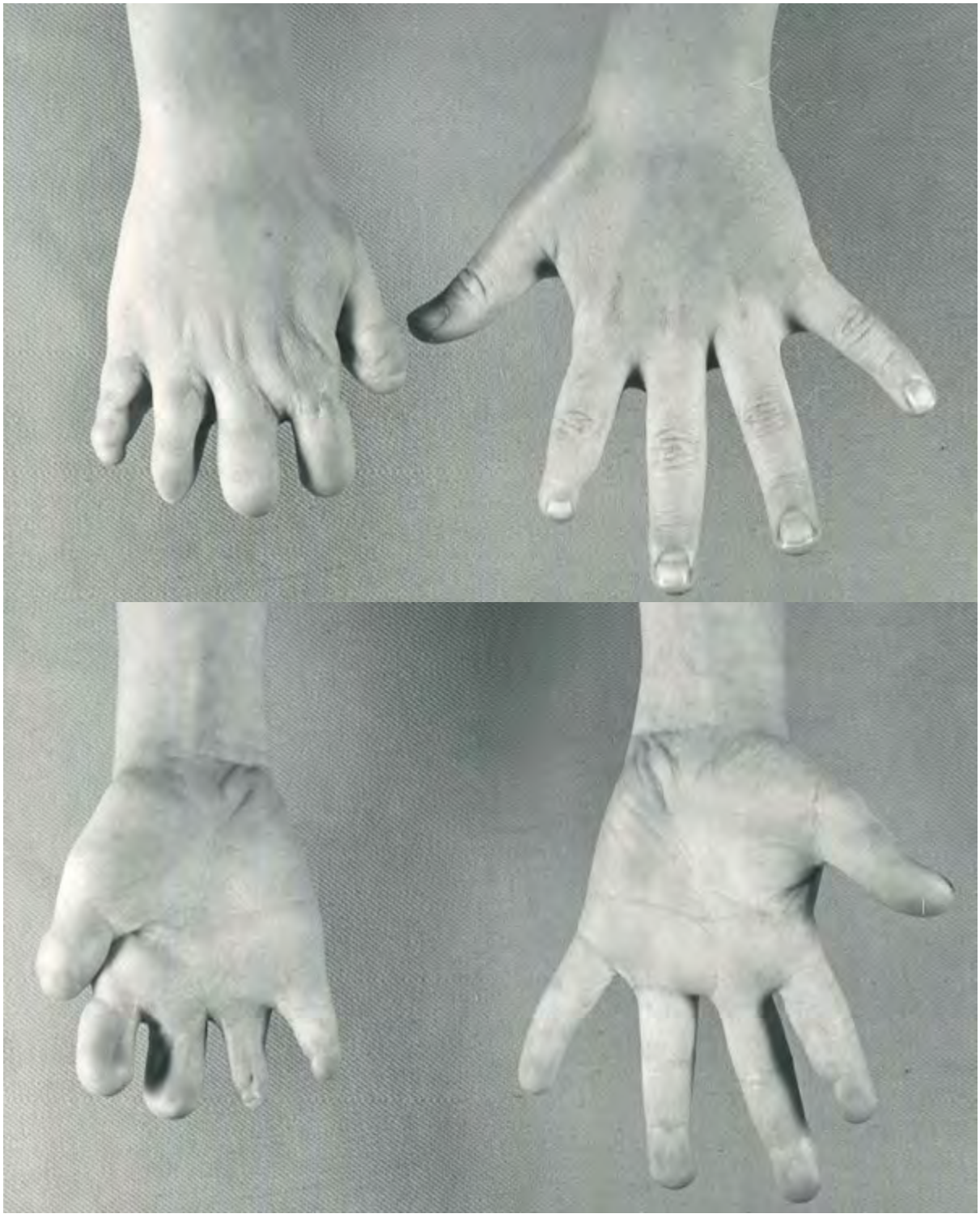
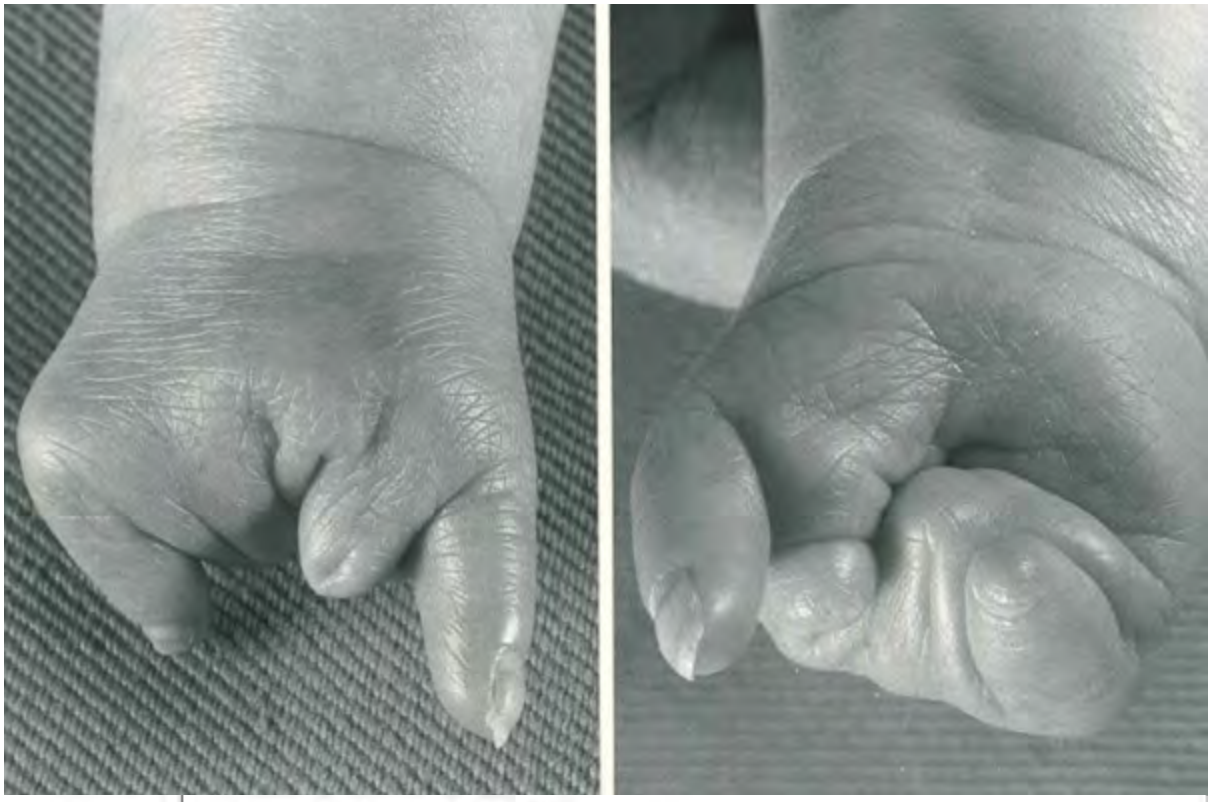


Figure 71: This is a case of constriction band sequence affecting the right hand causing auto-amputation of the more distal portions of the thumbs and the other fingers. He also underwent multiple z-plasties to improve the appearances of the digits.

CASE 46: *Constriction Ring Sequence*



14.8.87

SEENA T CLINIC BY MR LAMB

DIAGNOSIS :

HISTORY :

This boy has been born with the radial and ulnar components but absence of the central digits. There is a proximal phalanx of the index. At present the index metacarpal is not visible on Xray but I am sure would become so. He has tight thumb web. The easiest way to resolve this would be to resect the 2nd ray but in view of the proximal phalanx it would be better to remove the 3rd ray and mvoe the 2nd to that. The thumb appears to ahve no active IP mvoement. There are no flexion creases. The same appleis to the little finger where at present he flexes at MOJ.

No treatment required but should be seen in 3 months
I/T Mr Petrie cc GP.

DWL/MW

Figure 72: This is an interesting case in which the original diagnosis was uncertain. Mr Lamb described his deformity as "born with the radial and ulnar components, but absence of the central digits." The surgery that was considered at that time was to try and get improved opposition between the thumb and the little fingers as the central digits do not appear to be functioning well. The diagnosis here is probably that of constriction band syndrome with the central digits being joined at the tips (acrosyndactyly).

The Era of the Thalidomide Tragedy

THALIDOMIDE was marketed in 1957 by Chemie-Grunenthal as an effective drug in relieving morning sickness. Despite its wide distribution in Europe, the drug was refused its licensing in the United States. Subsequently there were growing suggestions that thalidomide was the cause of several birth defects, not just in Europe but also Canada, Japan and Australia. By 1961, the drug was withdrawn.

Several birth defects were associated with the teratogenic effects of thalidomide. With regards to the upper limb, this ranged from thumb polydactyly to complete phocomelia. There was a high incidence of radial dysplasia.

In the 1960s and 70s, several of these children presented to the Princess Margaret Rose Orthopaedic Hospital in Edinburgh. More and more national referrals were made to Douglas Lamb, and the hospital soon became a tertiary centre for the treatment of upper limb differences associated with thalidomide. Although tragic, this resulted in a rapid acceleration of experience gain in the treatment of certain conditions, like the radial longitudinal deficiency.

In 1977, Douglas published a landmark article on a continuing study of 68 patients with 117 club hands. (Lamb, 1977) He found that the anatomical findings and associated congenital abnormalities in the cases known to be related to thalidomide and in those in which the thalidomide was not a factor were similar, except that the incidence of other skeletal deficiency was higher in the thalidomide group. In this paper, he described the operation of centralisation which was described earlier.

In this section, we looked at some of the case files of children who were known to be affected by thalidomide. You will see the severity of the upper limb defects and also the creative use of a prosthesis to help with their function. Most of these patients will now be in their early 60s with age-related problems.

CASE 47: *Thalidomide*

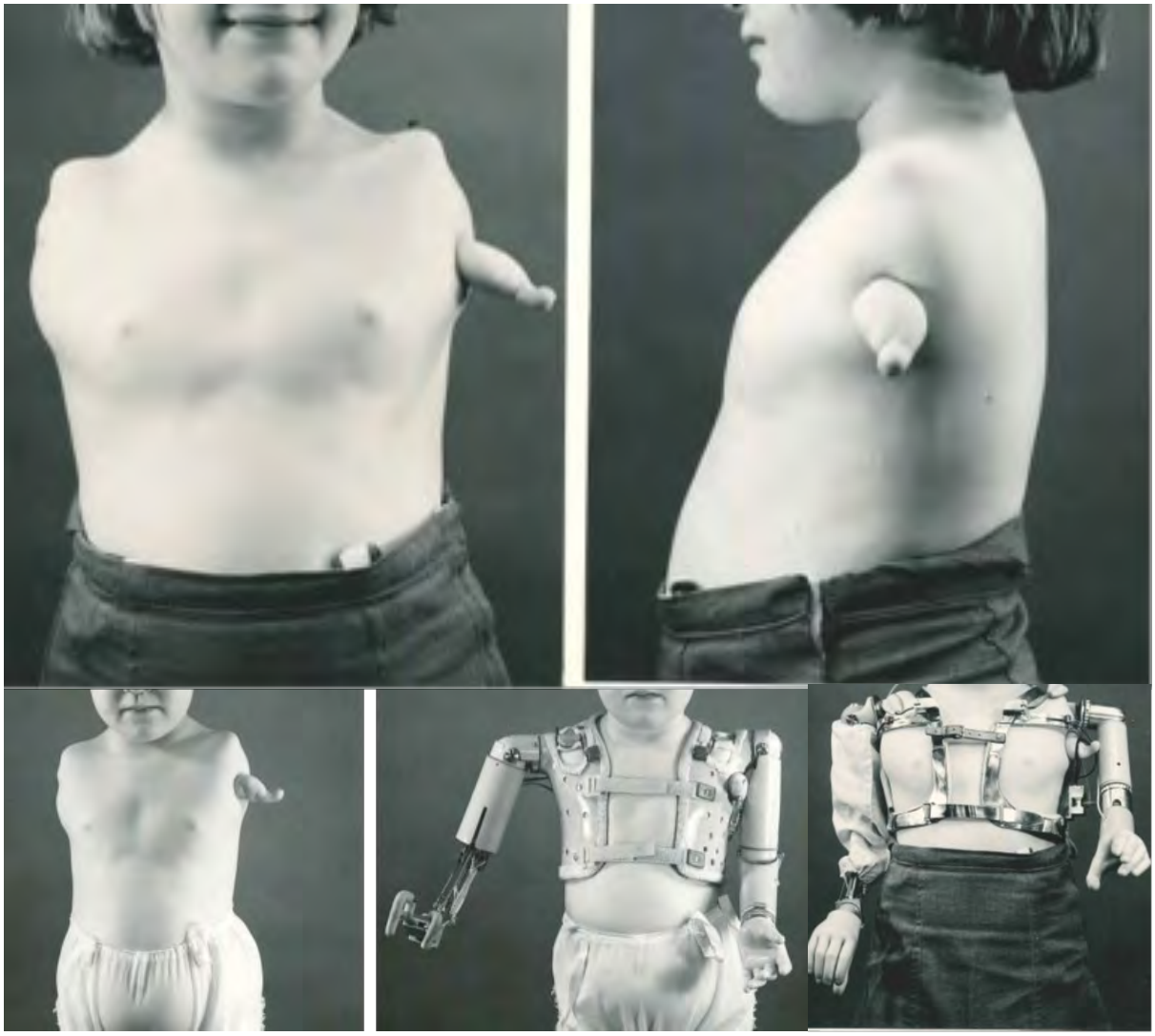


Figure 73: This patient has a complete amelia on the right side and a severe phocomelia-type condition on the left. She has been fitted with prosthetic limbs, but it is likely that in the future she will probably use her lower limbs for carrying out most of her functional activities, causing late problems with her hip joints.

CASE 48: *Thalidomide*

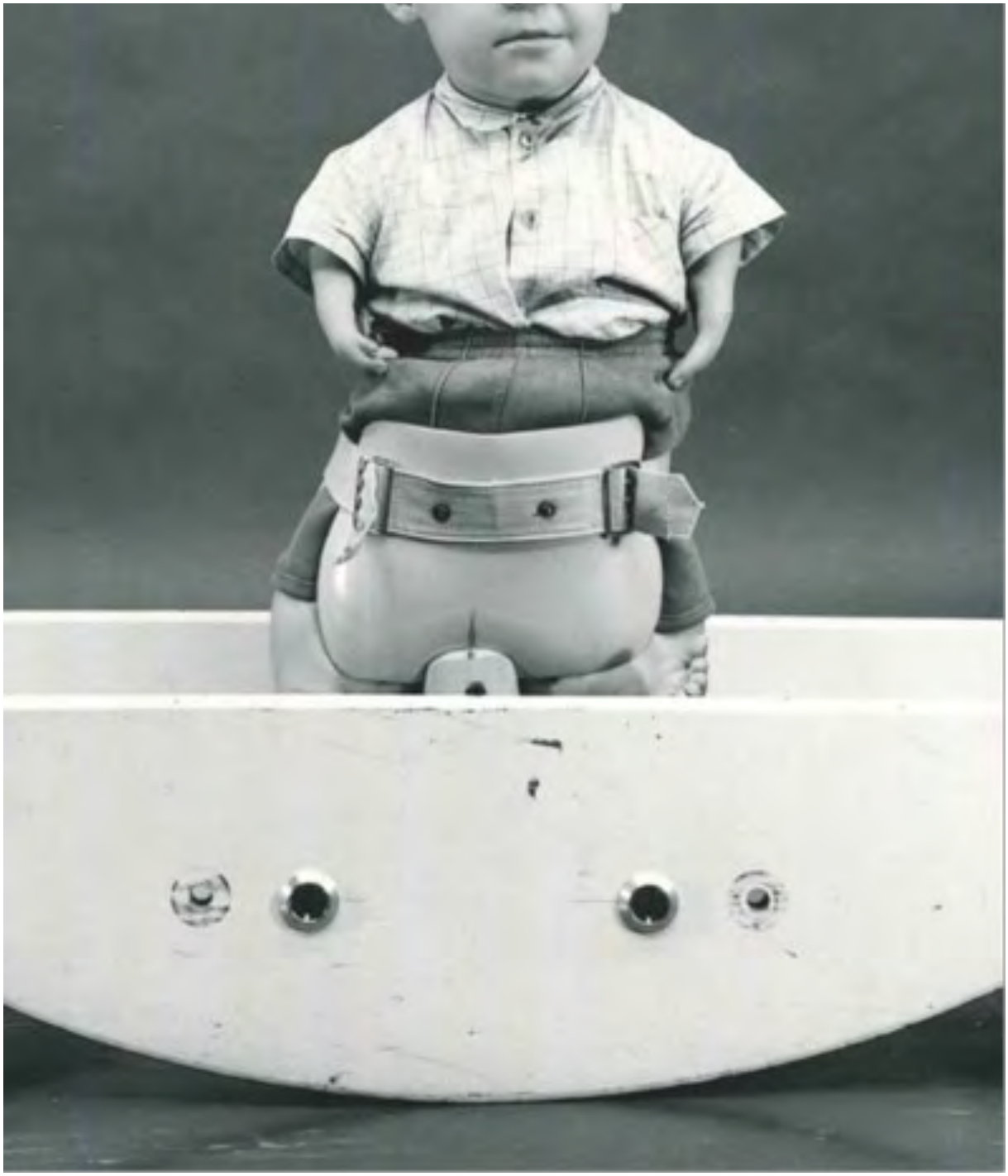


Figure 74: This patient has bilateral severe combined deficiencies of his upper limbs. He also has deficiencies of his lower limb and very limited walking ability.

CASE 49: *Thalidomide*

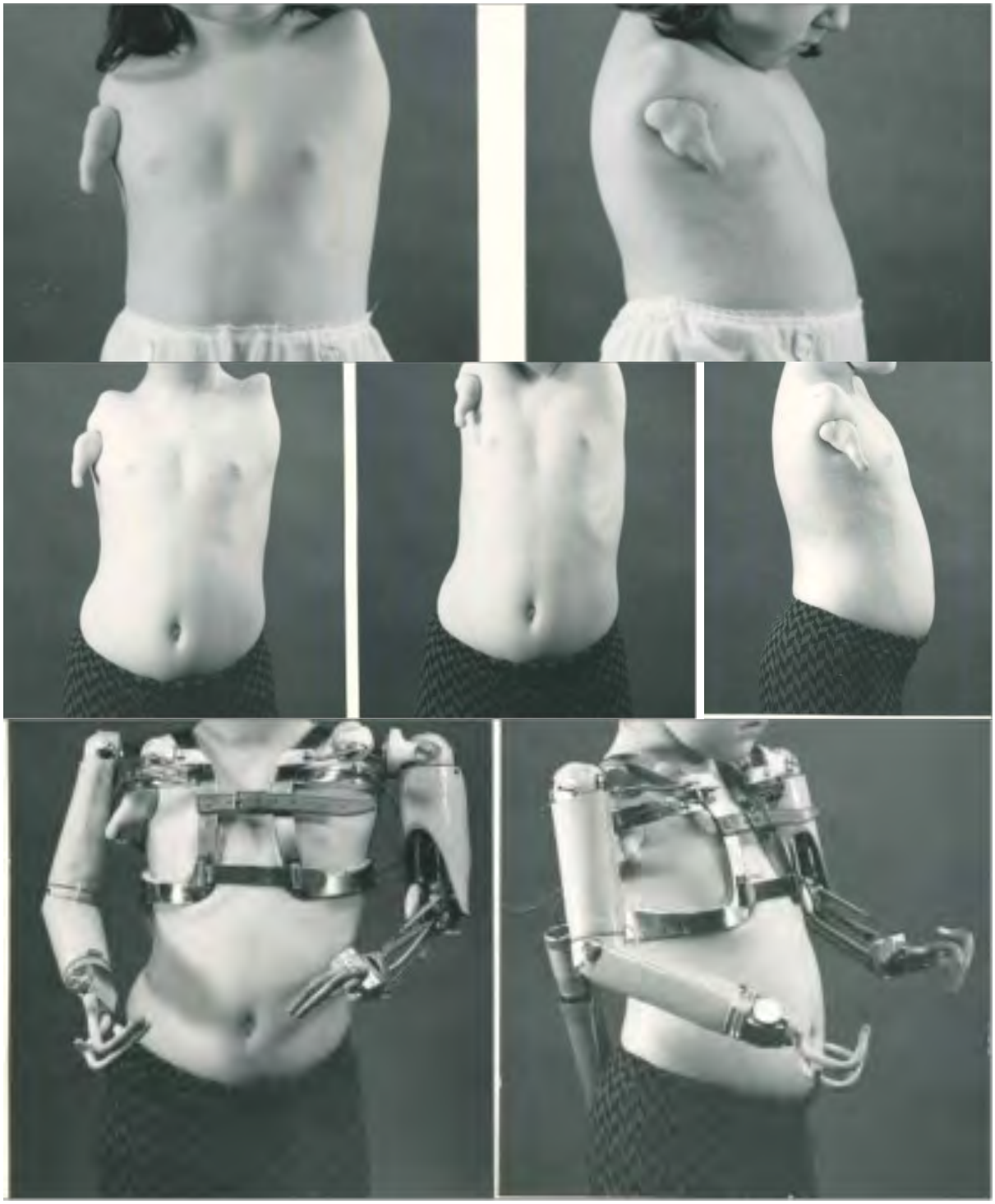


Figure 75: This patient has a severe phocomelia-type condition on the right and a complete amelia on the left. Again she has been fitted with prostheses to improve her upper limb function.

CASE 49 (continued): *Thalidomide*

Figure 76: Prostheses fitted for the above patient .

CASE 50: *Thalidomide*

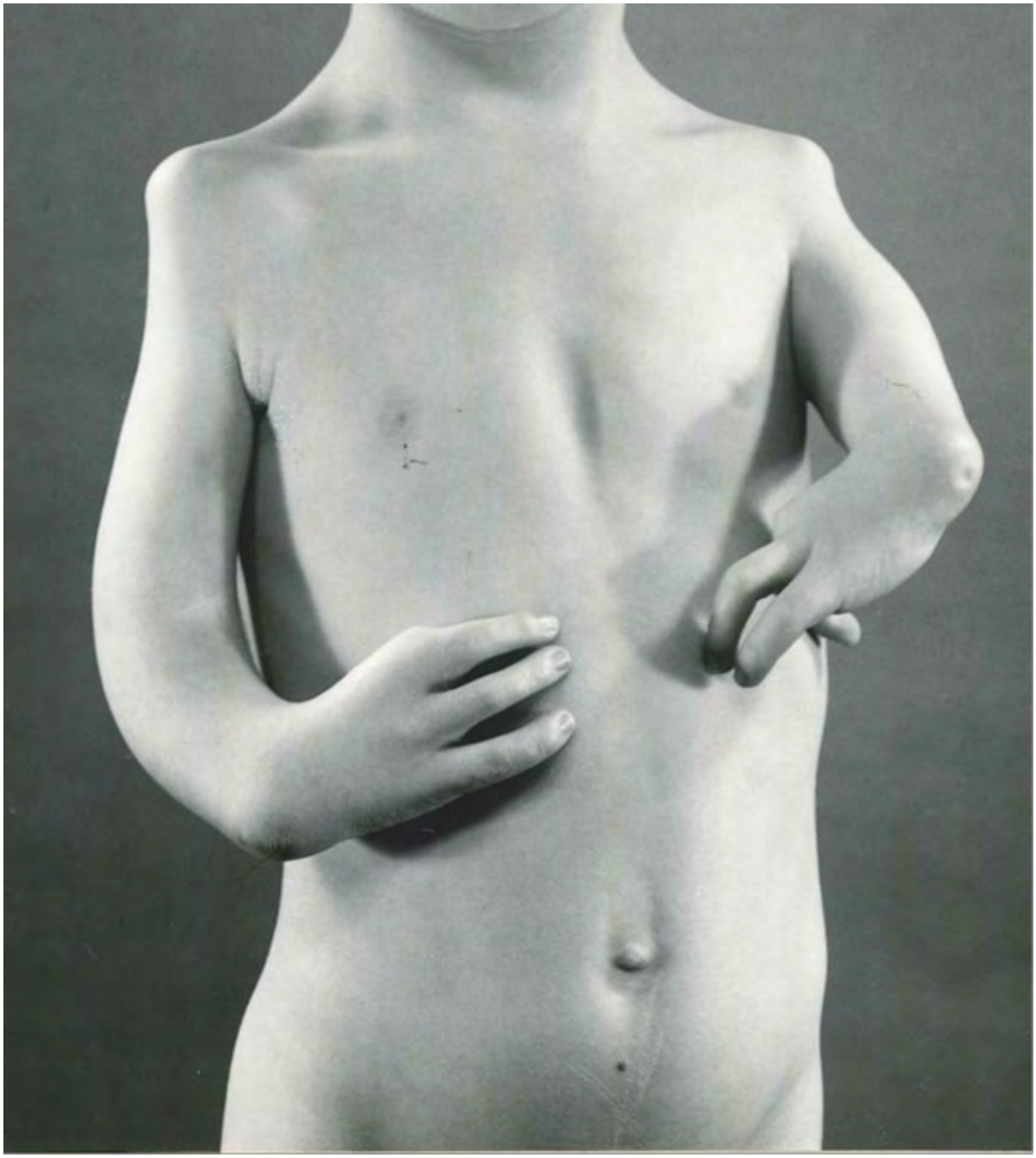


Figure 77: This patient has bilateral radial longitudinal deficiency.

CASE 50 (continued): *Thalidomide*

Figure 78: Thalidomide patient with bilateral radial longitudinal deficiency. On the left, the angulation is more severe and he has a complete thumb absence. On the right, despite the presence of a thumb, this is extremely hypoplastic and fused to the radial aspect of the index finger. He did not undergo surgery. You can see the detailed notes documenting the angulation of the upper limb as well as the various digit movements.

CASE 50 (continued): *Thalidomide*

CLINICAL HISTORY

6.3.67

ADMITTED: Self/Care Unit, P.M.R.:

Is a "thalidomide baby" with upper limb deficiencies. Is under the care of Mr. White at Law Hospital and has been admitted to the Thalidomide Self/Care Unit for toilet training and general assesament. Parents not interviewed.

O.E.:

Quick to settle. Intelligence and speech appear normal for age. Wears glasses with frosted left lense although eyesight with glasses apparently good.

No U.R.T. Infection.

No lymphadenopathy.

Teeth rotten.
No pharyngitis.
No cleft palate.

Chest: } Clear.
Lungs: }
Normal. No deformities.

C.V.S.:

P. 80 S.R. B.P.:

H.S.: 1 & 2 only. all areas.

Abdomen: NAD.

Lower Limbs:

Appear normal in all respects.

(L) Arm:

Uses Left as dominant hand.

Displacement. Humerus markedly short, with impaired shoulder range.

Elbow has 10 deg. flexion deformity and 30 deg. active flexion. Severe radial club hand deformity, with bursa and bunion over "heel" and marked tissue shortness on the radial side. Reduction deformity of digits. Has three normal looking fingers tho' index "broad based" and rudimentary jointed thumbapproaching from radial side of its proximal segment. Limited flexion of P.I.P. joint index. Other two digits appear normal. Sensation unimpaired.

(R) Arm:

Shoulder contours abnormal.
Humerus more normal length but marked reduction in shoulder movements.
Elbow 30 degs flexion deformity with 40 degs. further flexion.
Radial club hand deformity with bursa and bunion over "heel". Marked soft tissue shortness radial side.

Figure 79: Clinical notes of thalidomide patient with bilateral radial longitudinal deficiency.

CASE 50 (continued): *Thalidomide*

CLINICAL HISTORY—*Continuation Sheet*

6.3.67 Reduction deformity of hand with three digits and fairly normaly appearing thumb approaching from radial side.

P.I.P. joints of all three digits have grossly restricted flexion tho' in the index worse. Sensation normal.

FOR: X-ray of both upper limbs.

PGS/CST.

8.3.67 Seen on Ward round by Mr. Lamb:

We must obtain information from mother about this child's eyes, who he is attending and diagnosis.

I am not certain which is his dominant hand but his left seems better. No benefit could be obtained from surgery here.

Boy is completely untrained in self-care and I feel the parents co-operation should the sought for him to attend regularly - say every three months - for the time being. On discharge copy of letter to be sent to the Welfare Officer of the Thalidomide fund. PGS/CST.

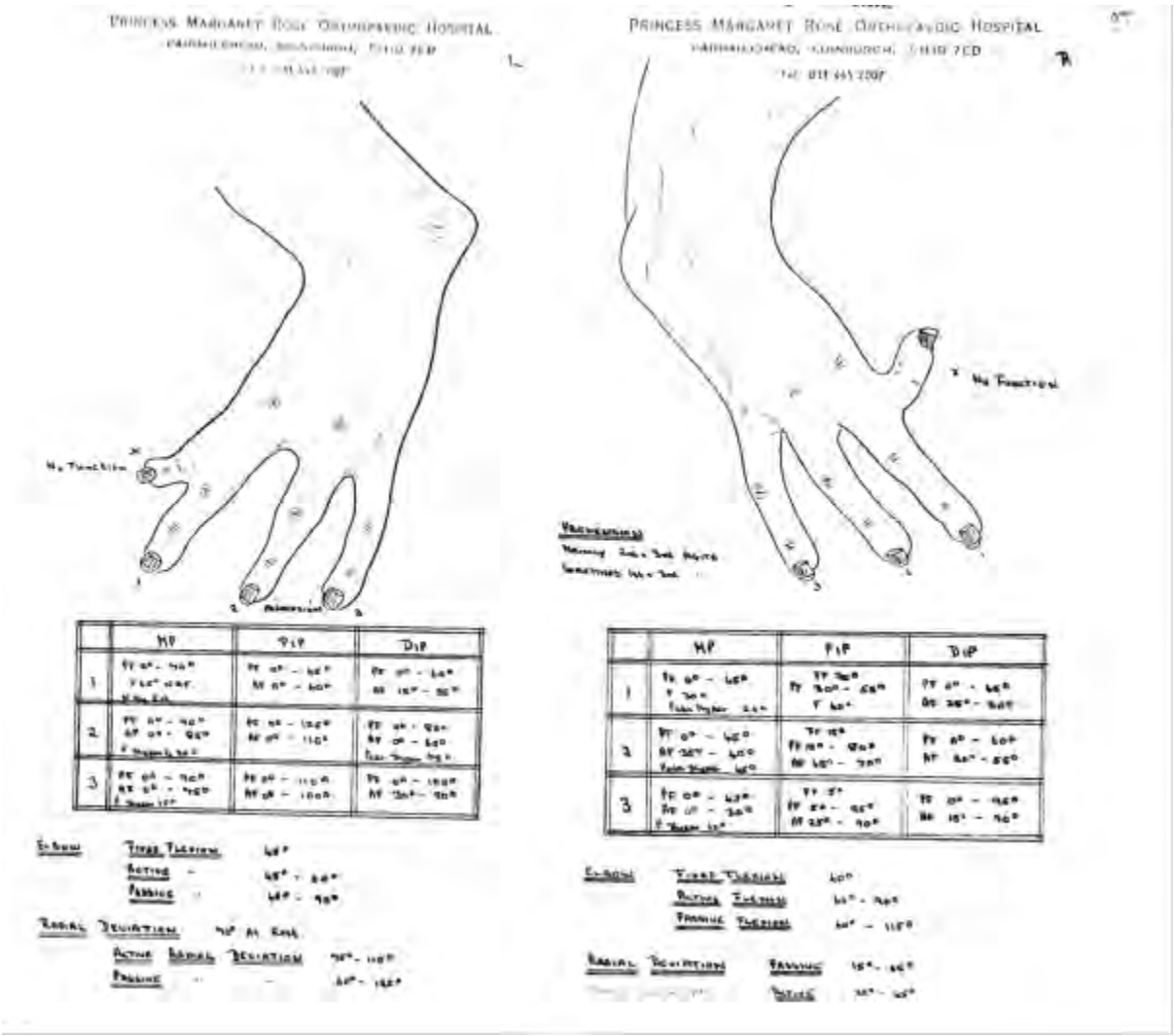


Figure 79 (continued): Clinical notes of thalidomide patient with bilateral radial longitudinal deficiency.

CASE 51: *Thalidomide*

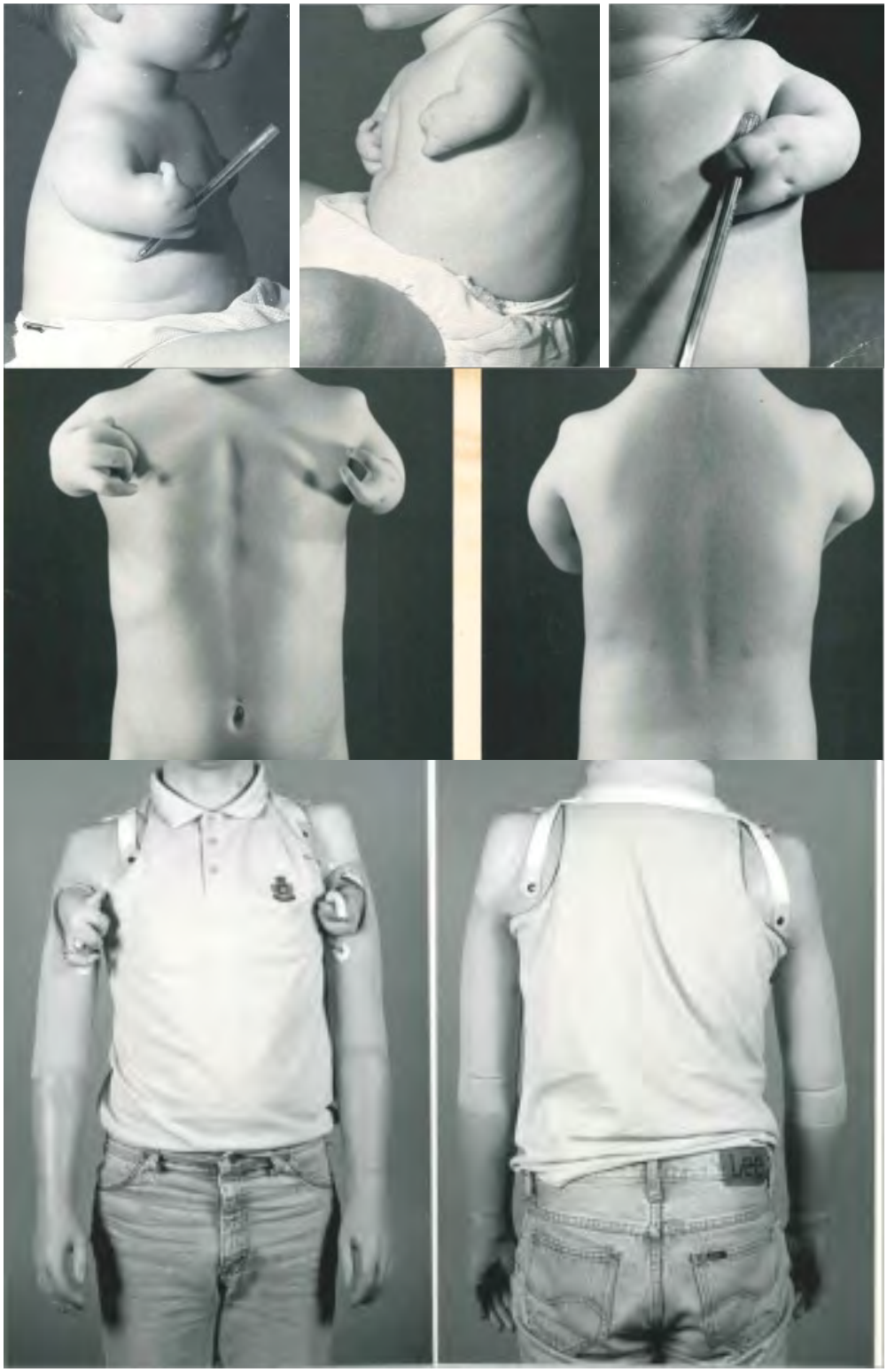


Figure 80: This is another patient with severe bilateral phocomelia-type conditions who was fitted with bilateral passive prostheses to fill the sleeves of clothing.

CASE 52: *Thalidomide*

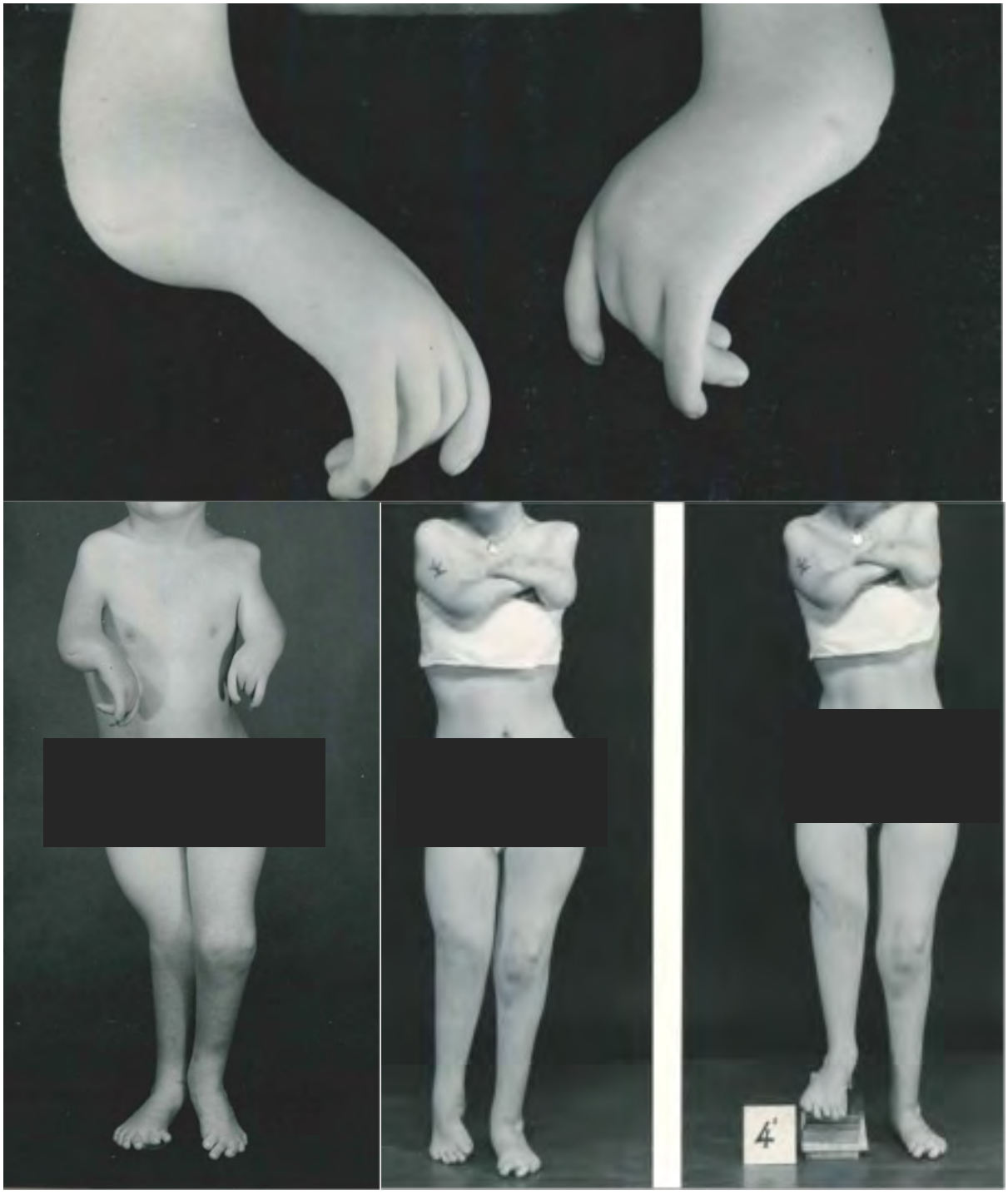


Figure 81: This is a patient with bilateral shortened upper limbs, which may correspond to either a proximal radial or ulnar dysplasia. She also has an underdeveloped tibia and other lower limb issues.

CASE 53: *Thalidomide*

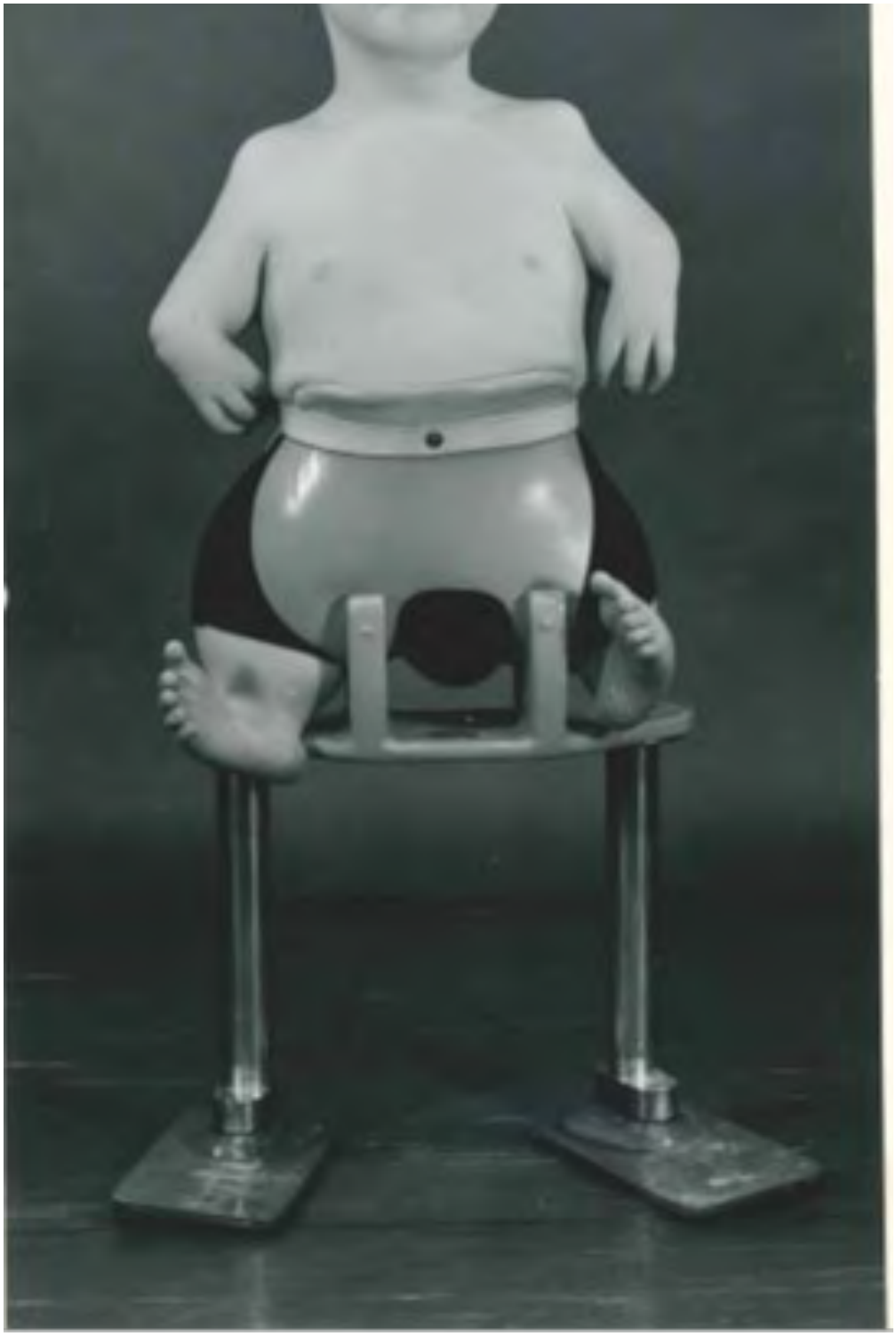


Figure 82: This is a patient with four-limb malformations as a result of thalidomide. All four limbs are severely shortened malformed limbs.

Final Thoughts

AFTER we scanned and catalogued these notes and photographs, they were taken to a storehouse at the University library. Further access can only be facilitated under strict Caldicott rules. I am encouraged that the notes were not destroyed as originally intended, but they are now stored safely.

While preparing this e-book, I found a document prepared by the national archives department of the government about the definition and purpose of archives: 'Archives are collections of documents or 'records' which have been selected for permanent preservation because of their value as evidence or as a source for historical or other research. Records are created by the activities of organisations and people; they serve an active purpose whilst in current use and some of them are later selected and preserved as part of an archival collection.'

This collection contains notes and pictures that are selected and preserved. In some ways, this has been a personal and sentimental journey but throughout the process, I was encouraged by those in the BSSH about the worthwhile nature of this exercise. The society is eager to honour Douglas Lamb and I hope to preserve his legacy, in which I believe that the contents of this e-book have achieved in some ways.

I also hope that the contents can be considered on their scientific merits. For those who see this as merely a historical exercise I hope that some insights provided will shed light on the evolution of care for children with congenital hand anomalies.

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