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

The treatment of longitudinal ulnar deficiency

R. J. LOVETT

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

Abstract

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

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

Introduction

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

Classification

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

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

Most classification schemes have emphasised the anatomic and radiologic abnormalities

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

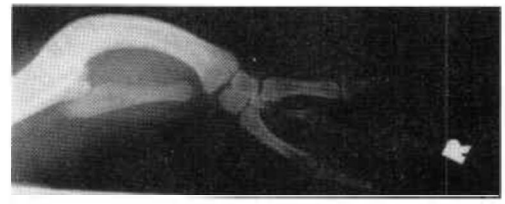

Fig. 1. A radiograph of longitudinal ulnar deficiency.

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

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

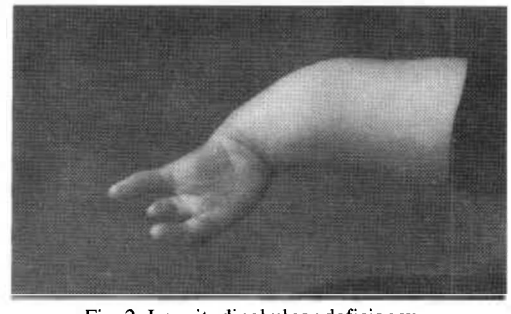

Fig. 2. Longitudinal ulnar deficiency.

Myths versus natural history

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

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

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

Table 1. Treatment of longitudinal ulnar deficiency.

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

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

REFERENCES

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

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

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

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

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

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

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

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

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

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

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