Hypoplasias and Aplasias of the Ulna
(Ulnar Club Hand)

14.1 Introduction

14.1.1 General Remarks

Ulnar dysplasias (synonyms: ulnar ray defect, ulnar club hand) are the result of an inhibitory malformation that mainly affects the ulnar parts of the upper limb. They are much less common than radial ray defects (Chap. 13). Usually one limb only is deformed. Sporadic appearance is well-known, heredity has been considered in some cases (see Kelikian 1974). Preponderance of the male sex has been suggested (Reimann-Hunzicker 1942).

A striking combination with dysplasias of the femur and fibula has been noted, the femur-fibula-ulna (FFU) syndrome (Kühne et al. 1967; Blauth 1967). Other combination defects are double thumbs (Kümml 1895; Kajon 1921) and aplasia of the tibia (Ke 1937; see also the experimental study by Degenhardt 1967). For further information concerning associated defects and also ulnar-fibular dysplasia, see Kelikian (1974).

Typical for an ulnar ray defect is shortening of the forearm, often with bowing in a dorsoradial direction. The hand deviates in an ulnar direction (manus valga, ulnar club hand). The deviation is caused partly by the bowing, partly by the insufficient or absent buttressing on its ulnar aspect.

Teratologic staging seems to be present, encompassing hypoplasias, partial aplasias, and total aplasias (see, among others, Bonola and Morelli 1972; other classification attempts: Pingel and Rompe 1971). In partial aplasias the defect is usually distally located. The ulna may be replaced by a strand of tight connective tissue.

In general there is some shortening of the radius. It too is bowed in a dorsoradial direction. The dysplastic head of the radius is frequently displaced, especially when the ulna is hypoplastic. In such a case it can be palpated lateral to the lateral humeral epicondyle. Synostoses between humerus and the hypoplastic radius, either in a position of flexion or of extension, are likewise well-known; radiologically it may be impossible to distinguish between humeral and radial elements. Cases of bizarre bifurcation of the distal humerus have been reported. They seem to be the result of a synostosis between humerus and ulna. Longitudinal growth of the humerus is frequently impaired too, particularly in the presence of more severe types of ulnar dysplasia.

Hypo- and aplasias of ulnar ray carpal bones are common (os triquetrum, os pisiforme, os hamatum, and occasionally os capitatum). Synostoses of extant carpal bones, (in addition and simultaneously), have been described.

Aplasia of the ulnar digits (oligodactyly) is nearly always observed. The ring and little fingers are most frequently missing, absence of the Vth ray alone is
rare. Concomitant differentiation anomalies of the radial border ray may be present, i.e., thumb hypoplasia with partial syndactyly. Also fully developed fingers are sometimes misshapen; cutaneous and osseous syndactylies, spoon hands, brachydactylies, joint aplasias, finger duplications, camptodactylies and other anomalies have been reported. In extreme cases of ray reduction only one tri- or biphalangeal finger may be present (monodactyly).

Soft tissue anomalies, especially of the ulnar part, are found in addition to the bony changes. The literature (see Wierzejewski 1910) contains reports of aplasias and insertion anomalies of m. triceps, m. biceps, and the ulnar wrist joint flexors and extensors, as well as aplasias of m. palmaris longus, m. extensor digiti quinti, m. supinator, and m. anconeus. The ulnar nerve and artery are frequently missing or take an abnormal course.

14.1.2 Indications for Surgery

Functional disability is quite severe in most cases of ulnar dysplasia. Surgery on the other hand is indicated only if marked improvement of function may be expected. Surgery may be considered in the presence of the following deformities:

- **Aplasias of the elbow joint** with the joint in extension or hyperextension or in marked flexion. The most satisfactory position of function is an angle of about 90°, (depending on the amount of shortening of the forearm). It can be obtained with an angulation osteotomy.

- **Radial head dislocations** may result in marked limitation of motion or malposition. The protruding head can be a source of local irritation. Therapeutically it may be necessary to resect the radial head.

- **Shortening of the ulna and bowing of the radius** with ulnar deviation of the hand require surgery if the axial deviation results in markedly impaired function and if neutral position of the hand may be obtained by corrective osteotomy of the radius, or if by an (additional) lengthening osteotomy of the ulna buttressing of the hand can be improved, or by a radius-pro-ulna operation (see Bonola and Morelli 1972; Kelikian 1974). A derotation osteotomy may be required in cases of fixed supination of the forearm.

- **Malformations of the hand** if marked functional improvement can be expected by correction of partial finger duplications or of syndactylies, or by creation of a post in cases of monodactyly, if a metacarpus is present (see also Chaps. 2, 4, 8.7).