Ulnar deficiency

Twenty-nine patients with 34 ulnar deficient limbs were studied; the average follow-up was 7.7 years. To permit early determination of prognosis and decisions regarding treatment, four subtypes of this condition have been identified. Progressive ulnar deviation of the wrist was not observed in any case, and the fibrocartilaginous ulnar anlage was resected only to correct fixed deformities of the wrist. The most useful operation was humeral or radial osteotomy to correct the hand on flank deformity. A group of patients who were optimally treated with prosthetic fitting is discussed. (J HAND SURG 11A:822-9, 1986.)


Longitudinal deficiency of the ulna, complete or partial, is a rare affection of the upper limbs; radial deficiency occurs ten times more frequently. Ulnar deficiency did not occur in Entin’s series of 61 cases of upper limb deficiency, or in the series of Henkel and Willert of 557 malformed upper limbs. Accordingly, most series of patients with ulnar deficiency have been relatively small. Several different classification systems based primarily on radiographic features have been proposed. Kummel defined three types on the basis of the radiographic appearance of the elbow joint as follows: type A, normal appearing joint; type B, radiohumeral synostosis; type C, dislocated radial head. Ogden and associates proposed a system based upon the degree of ulna visible radiologically. Each of these systems adequately classifies the deformities, yet they are not useful in making clinical decisions regarding prognosis and treatment. We present a classification system on the basis of our long-term follow-up of a large number (29) of patients with longitudinal deficiency of the ulna. This system is useful for formulating a prognosis and planning treatment in this condition.

Materials and methods

Twenty-nine patients with 34 ulnar deficient limbs were studied. Multiple clinical and radiographic features were retrospectively reviewed to determine the influence of each one, and the combinations of different ones, on the overall prognosis and on the clinical decision making process.

The patients were seen at the Shriners Hospital, Springfield Unit, over a 30-year period between 1954 and 1984. Follow-up averaged 7.7 years, with a range of 2 months to 16 years, 8 months. In 19 cases the left side only was affected, in five cases the right side only was affected, and five cases were bilateral. There were 18 male patients and 11 female patients. There was no family history of a similar deformity in any of the cases.
Classification of elbow and forearm deformities

The limbs showed four distinct subtypes by radiographic and clinical examinations. They are as follows:

Type A. (14 limbs) Type A has a dislocated radial head, with a variable degree of bowing of the radius. Ossification of the ulna is delayed in most cases, but progressive ossification is seen in serial x-ray films. Most patients lack normal elbow flexion/extension and forearm pronation/supination. The elbow has a variable range of motion, but it tends to rest in an average of forty° of flexion. No patient has a cubital web. The limited range of motion does not prove to be a major functional deficit. With compensatory shoulder motion, all patients can adequately position their hand in space. Elbow instability is not a complaint in any of these patients (Figs. 1 and 2).

Type B. (3 limbs) In Type B deformity the radial head is dislocated, there is complete and persisting absence of the ulna, and there is cubital webbing. The average elbow flexion deformity measured 125° and all were greater than 110°. All three of these patients had limited ability to position and use the hand in front of
Fig. 4. The elbow of the patient in Fig. 3. Nearing skeletal maturity, the features include complete absence of the ulna, dislocated radial head, and marked flexion of the forearm segment.

The body because of the flexed elbow. These patients use their ulnar deficient limbs to help perform tasks about the head and chest only (Figs. 3 and 4).

Type C. (14 limbs) Type C patients have a radiohumeral synostosis, with a variable amount of ulna. As with type A patients, ossification of the ulna is delayed, but it always eventually appears. The angle between the humerus and radius is variable, measuring between 15° and 100° in our patients. The radius is bowed, but in no case did the bowing progress with time (Fig. 5).

The "hand on flank" deformity was present in nine of fourteen limbs. This deformity consists of a hyperpronated forearm, bowing of the radius, and flexion and rotation of the elbow so that the hand faces posteriorly and tends to lie on the flank or buttock at rest (Fig. 6, A). To place the hand in a functional position in front of the plane of the body, the patient externally rotates at the shoulder (Fig. 6, B).

Type D. (3 limbs) Type D deformity shows a radiohumeral synostosis, with a straight radius and an ulna of near normal length but of markedly diminished diameter. The forearm is in neutral rotation. The elbow is at an average of 45° of flexion. With the straight radius, no patient exhibits the "hand on flank" deformity. These patients function well, and they can easily position the hand in front of the body without excessive external rotation at the shoulder (Fig. 7).

Associated deformities

Wrist. The wrist was ulnarly deviated to a significant degree in 14 of the 34 cases. The ulnar deviation was not progressive in any of the cases.

Hand. A three-digit hand was the most common pattern seen. Nineteen of the 34 cases (56%) showed this variation (Table I).

The thumb was absent in 7 cases, hypoplastic in 7 cases, and of normal size in 20 cases. It was frequently of normal size but had no opposition power. In these cases the first webspace was contracted.

Syndactyly, always simple, was present in 13 of the hands. The most frequently observed hand anomaly was fixed flexion deformity of the interphalangeal joints. This was present in 23 of the cases.

Shoulder. There were four cases of ipsilateral shoulder instability. These patients could either subluxate or dislocate their shoulders. In two instances this occurred while the patient maneuvered the hand into a functional position (cases 19 and 23). The dislocations were easily reduced by the patients and all patients were asymptomatic.

Miscellaneous. Nineteen of the 29 patients had anomalies remote from the ulnar deficient limbs. Eighteen had additional musculoskeletal anomalies—five with toe syndactyly, three with contralateral above el-
bow amputations, three with bilateral fibular deficiency, two with bilateral proximal focal femoral deficiency, four with scoliosis, and three with adactylyia of otherwise normal limbs. Also represented were congenital dislocations of the hip, congenital coxa vara, congenitally short femur, torticollis, and two patients with absence of the ipsilateral pectoralis major. The central nervous system was involved in one case of Sturge-Weber syndrome, and in one case of grand mal seizures. Also present was one case each of hypospadius, bicornuate uterus, branchial cyst, and congenital diaphragmatic hernia.

Nonoperative treatments. Most patients were evaluated at an early age. They were started on a program of passive stretching of the ulnarly deviated wrist and night splinting of the wrist. The patients were encouraged to perform two-handed activities and to use their deficient limbs as much as possible. Two patients with type B deformity were fitted with above elbow type prostheses over the flexed elbow. One patient wore the prosthesis full time, with greatly improved overall function (case 17). The other patient rejected his prosthesis after being fit with it at age ten (case 16).

Operative treatment of elbow and forearm deformities. Of the twenty type A, B, and D limbs, only one patient was treated surgically for the elbow or forearm deformity. This procedure was an attempted release of a cubital web; the operation was unsuccessful (case 15). The type A patients had enough mobility of the elbow and forearm to position the hand in front of the body. Type D patients, because of the straight forearm and despite the fixed elbow, could position their hands adequately with compensatory shoulder motion.

Of the fourteen type C limbs, five had a total of eight
Table I

<table>
<thead>
<tr>
<th>Patient</th>
<th>Type</th>
<th>Extremity</th>
<th>Digits</th>
<th>Follow-up</th>
<th>Operations/prostheses</th>
<th>Associated anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>A</td>
<td>L</td>
<td>4</td>
<td>3 mo</td>
<td>None</td>
<td>Camptodactyly; Sturge-Weber syndrome</td>
</tr>
<tr>
<td>2</td>
<td>A</td>
<td>L</td>
<td>3</td>
<td>7 mo</td>
<td>Syndactyly release</td>
<td>Hypospadia; syndactyly of toes</td>
</tr>
<tr>
<td>3</td>
<td>A</td>
<td>R</td>
<td>3</td>
<td>2 yr</td>
<td>Release constriction band at wrist</td>
<td>Left hypoplastic humerus, dislocated elbow, adductia 4th, 5th fingers; bilateral absent 5th rays of feet; bilateral genu valgus</td>
</tr>
<tr>
<td>4</td>
<td>A</td>
<td>R</td>
<td>3</td>
<td>10 yr, 3 mo</td>
<td>None</td>
<td>Syndactyly, left 2nd + 3rd, 4th + 5th toes; 1½° leg length discrepancy</td>
</tr>
<tr>
<td>5</td>
<td>D</td>
<td>L</td>
<td>4</td>
<td></td>
<td>None</td>
<td>Congenital short femur, left, syndactyly left 4th + 5th toes; left congenital BE amputation</td>
</tr>
<tr>
<td>6</td>
<td>A</td>
<td>L</td>
<td>2</td>
<td>2 yr, 10 mo</td>
<td>Syndactyly release, rotational osteotomy of radial digit</td>
<td>Absent right 5th ray</td>
</tr>
<tr>
<td>7</td>
<td>A</td>
<td>L</td>
<td>1</td>
<td>15 yr</td>
<td>Opposition post</td>
<td>Right congenital AE amputation; bilateral PFFD with fibular deficiency</td>
</tr>
<tr>
<td>8</td>
<td>A</td>
<td>L</td>
<td>2</td>
<td>16 yr</td>
<td>Syndactyly release</td>
<td>Left congenital coxa vara; left clubfoot; bilateral genu valgus</td>
</tr>
<tr>
<td>9</td>
<td>A</td>
<td>L</td>
<td>3</td>
<td>2 yr, 6 mo</td>
<td>Pollicization radial border digit</td>
<td>Infantile idiopathic scoliosis</td>
</tr>
<tr>
<td>10</td>
<td>A</td>
<td>L</td>
<td>5</td>
<td>2 yr, 6 mo</td>
<td>Excision fibrocartilaginous anlage, lengthening FCU; syndactyly release</td>
<td>Syndactyly left 2nd through 5th toes</td>
</tr>
<tr>
<td>11</td>
<td>A</td>
<td>L</td>
<td>3</td>
<td>1 yr, 11 mo</td>
<td>None</td>
<td>Absent left pectoralis major; bilateral pes planus</td>
</tr>
<tr>
<td>12</td>
<td>A</td>
<td>L</td>
<td>3</td>
<td>14 yr, 3 mo</td>
<td>Syndactyly release, correction of ulnar deviation with fusion of wrist</td>
<td>Idiopathic scoliosis</td>
</tr>
<tr>
<td>13</td>
<td>A</td>
<td>R</td>
<td>3</td>
<td>11 yr, 2 mo</td>
<td>None</td>
<td>Bilateral PFFD; Right congenital AE amputation; Right equinovalgus foot</td>
</tr>
<tr>
<td>14</td>
<td>A</td>
<td>L</td>
<td>3</td>
<td>2 mo</td>
<td>None</td>
<td>Idiopathic scoliosis</td>
</tr>
<tr>
<td>15</td>
<td>B</td>
<td>R</td>
<td>3</td>
<td>14 yr, 9 mo</td>
<td>BE prosthesis</td>
<td>None</td>
</tr>
<tr>
<td>16</td>
<td>B</td>
<td>L</td>
<td>2</td>
<td>1 yr, 9 mo</td>
<td>Release of cubital web, rotational osteotomy ulnar metacarpal</td>
<td>Absence of right pectoralis major</td>
</tr>
</tbody>
</table>

The objective of the operation is to place the hand in front of the trunk, with the forearm held in midrotation. Approximately 60° of elbow flexion is sought.

Radiographs are evaluated (1) to confirm the presence of elbow and/or forearm synostosis and (2) to determine the optimal site for osteotomy.

The patient is placed in a supine position on the operating table. A lateral incision is used and the osteotomy site is exposed subperiosteally. Marks are made on the bone to verify its preosteyotomy orientation. A closing wedge osteotomy is done, with its base placed anteriorly and medially (on the basis of the resting position of the limb) and the apex posteriorly and laterally. If necessary, an added segment of bone (0.5 cm to 1.0 cm) can be removed to lessen the tension on the soft tissues. The fragments are fixed with crossed wires. The incision is closed, and a cast is applied to the limb.

Because it is difficult to achieve the desired position...
<table>
<thead>
<tr>
<th>Patient</th>
<th>Type</th>
<th>Extremity</th>
<th>Digits</th>
<th>Follow-up</th>
<th>Operations/prostheses</th>
<th>Associated anomalies</th>
</tr>
</thead>
<tbody>
<tr>
<td>17</td>
<td>B</td>
<td>L</td>
<td>1</td>
<td>3 yr, 3 mo</td>
<td>Fit with elbow disarticulation prosthesis</td>
<td>Central ray deficiency right hand; syndactyly 1st and 2nd, 3rd and 4th toes, right; idiopathic scoliosis; brachial cyst; bipartite uterus</td>
</tr>
<tr>
<td>18</td>
<td>C</td>
<td>L</td>
<td>2</td>
<td>14 yr, 3 mo</td>
<td>None</td>
<td>Posterior subluxation of left shoulder</td>
</tr>
<tr>
<td>19</td>
<td>C</td>
<td>L</td>
<td>5</td>
<td>15 yr, 6 mo</td>
<td>Distal humeral osteotomy; distal humeral osteotomy revision</td>
<td>None</td>
</tr>
<tr>
<td>20</td>
<td>C</td>
<td>R</td>
<td>2</td>
<td>9 yr, 9 mo</td>
<td>Syndactyly release</td>
<td>None</td>
</tr>
<tr>
<td>21</td>
<td>C</td>
<td>L</td>
<td>3</td>
<td>9 yr, 11 mo</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>22</td>
<td>C</td>
<td>R</td>
<td>3</td>
<td>3 mo</td>
<td>Ulnar wrist release, radial osteotomy; syndactyly release</td>
<td>None</td>
</tr>
<tr>
<td>23</td>
<td>C</td>
<td>L</td>
<td>5</td>
<td>8 yr, 2 mo</td>
<td>Humeral osteotomy; Radial osteotomy</td>
<td>Voluntary ant. shoulder dislocation; congenital diaphragmatic hernia</td>
</tr>
<tr>
<td>24</td>
<td>C</td>
<td>R</td>
<td>3</td>
<td>4 yr, 5 mo</td>
<td>Syndactyly release; Radial osteotomy, excision of ulnar anlage</td>
<td>Torticollis</td>
</tr>
<tr>
<td>25</td>
<td>C</td>
<td>R</td>
<td>3</td>
<td>14 yr, 10 mo</td>
<td>First web space deepening; rotational osteotomy of thumb metacarpal</td>
<td>Grand mal seizures</td>
</tr>
<tr>
<td>26</td>
<td>C</td>
<td>L</td>
<td>3</td>
<td>7 mo</td>
<td>Middle ray resection; radial osteotomy</td>
<td>None</td>
</tr>
<tr>
<td>27</td>
<td>C</td>
<td>L</td>
<td>3</td>
<td>5 yr, 5 mo</td>
<td>Syndactyly release</td>
<td>None</td>
</tr>
<tr>
<td>28</td>
<td>C</td>
<td>R</td>
<td>3</td>
<td>None</td>
<td>None</td>
<td>None</td>
</tr>
<tr>
<td>29</td>
<td>D</td>
<td>L</td>
<td>2</td>
<td>16 yr, 8 mo</td>
<td>Syndactyly release; fitting with opposition post</td>
<td>Right congenital AE amputation; Bilateral fibular deficiency</td>
</tr>
</tbody>
</table>

Precisely, it is not unusual to need a second osteotomy for further correction. This is generally performed 1 to 3 years later.

**Operative treatment of associated deformities.**

**Wrist.** Five of the 34 limbs were treated with resection of the ulnar anlage and ulnar wrist release. One patient also had a wrist arthrodesis. Operations were performed for significant ulnar deviation of the wrist, not for progressive deformity, as this was not observed in any patient. The fibrocartilaginous ulnar anlage was not resected in the remaining cases since its presence was not found to be associated with either progressive bowing of the radius or proximal migration of the radial head.

**Hand.** Three patient’s had osteotomy of the thumb metacarpal. Two had simultaneous deepening of the first webspace. All three had excellent results. One patient was treated with a rotational osteotomy of the ulnar border digit since the thumb was hypoplastic and poorly motored. Ten syndactyly releases were performed with good results. Camptodactyly was not surgically treated.

**Discussion**

The major upper limb functional deficits in patients with ulnar deficiency relate to the elbow and forearm deformities. Several reconstructive procedures have been advocated for the elbow and forearm in ulnar deficiency. Goddu described a procedure for reconstruction of the distal ulna and the radiohumeral articulation. Vitale first described the use of the one bone forearm technique of Hey-Groves for ulnar deficiency. Straub modified this technique by resecting the fibrocartilaginous ulnar anlage at the time of construction of the one bone forearm. Other authors adopted this technique and proposed that it could be used to...
provide stability to the forearm and to correct a supination deformity. Laurin and Farmer observed good function in their patients without this procedure and did not recommend it. We chose not to construct a one bone forearm in any of our type A patients (who would be candidates for such procedure according to the above authors) for two reasons. First, progressive forearm shortening caused by the dislocated radial head being driven proximally did not occur. Second, many of our type A patients retained a useful arc of forearm rotation, as Blair noted in his patients. Such motion would be eliminated by the one bone forearm operation.

Frantz and O'Rahilly recommended above elbow prosthetic fitting for patients with ulnar deficiency and an acutely flexed elbow with a cubital web. Their results with release of the cubital web were poor. These patients would be classified as type B in our system. Our experience with these patients has been similar, with one unsuccessful attempt at release of a cubital web, and one successful above elbow prosthesis wearer. Fitting a prosthesis over a sensate limb requires early acceptance by the patient and family. This explains why one patient rejected the prosthesis when first fitted with it at age ten.

It is essential that type C patients be differentiated from type D patients. The former may require corrective osteotomies if the combined position of the elbow, forearm, and wrist is such to yield the “hand on flank” deformity. The type D patients should not require an operation other than for their hands. The two types can be distinguished at an early age, and it is helpful to offer this prognostic information to the parents.

The fibrocartilaginous ulnar anlage has been described as the anatomic structure responsible for progressive bowing of the radius, dislocation or synostosis of the radiohumeral joint, ulnar subluxation of the wrist, and loss of longitudinal growth of the radius. Ogden and colleagues recommended treatment by early resection to prevent these problems, and they reported improvement in the bowing of the radius after this procedure. However, Broudy and Smith did not note increasing ulnar deviation of the wrist or late dislocation of the radial head in their series of 26 cases. We did not observe any of the previous problems in 29 limbs that did not have the ulnar anlage resected. We found this procedure useful to correct the ulnarly deviated wrist. We do not recommend its use to prevent progressive bowing of the radius, late radial head dislocation, or loss of longitudinal growth of the radius.

The hand deformities are best treated in the usual manner. The most useful procedures were rotational osteotomy of a border digit and syndactyly release. The high incidence of flexion deformities of the interphalangeal joints has not been previously reported.

Atrophy and weakness about the shoulder girdle have been noted by others. Our description of four patients with shoulder instability is a previously unreported finding. This represents another feature of the generalized limb bud defect. The finding is of clinical significance if the patient is dislocating the shoulder in order to put the hand into a functional position. This can be treated, as was done in two of our patients, through corrective osteotomies to put the hand in a position of function, while allowing the shoulder to remain in neutral position.

Our series of patients had a very high percentage of associated anomalies. This may be because of our status as a referral center for limb deficient children and to the special interest in this problem of the senior author (L. M. K.).

Conclusions

1. We believe this system for the classification of ulnar deficiency is useful for predicting prognosis and planning treatment in a given case.

**Type A. Dislocated radial head with bowed radius.** These patients do not require osteotomy to position the hand in front of the body. One bone forearm construction is not necessary since progression of the radial bow, elbow instability, and loss of forearm length caused by the dislocation have not been observed.

**Type B. Dislocated radial head, straight radius, cubital web with flexed elbow.**

Function is improved by early fitting with an above elbow type prosthesis. Release of the web is not successful.

**Type C. Radiohumeral synostosis with bowed radius, variable presence of ulna.**

Patients in this group, with more severe combinations of deformities, will benefit from radial or humeral osteotomy to position the hand in front of the body.

**Type D. Radiohumeral synostosis with straight radius, diminutive ulna.**

These patients are easily identified at birth and should require no treatment for their elbow and forearm deformities.

2. Resection of the fibrocartilaginous ulnar anlage was useful only to correct fixed ulnar deviation at the wrist.

3. A new association is described—shoulder instability in the patient with ulnar deficiency.
REFERENCES


Brachymetacarpia and brachyphalangia

The x-ray features of 325 patients, with shortening of the metacarpals and/or phalanges were analyzed. Most patients with congenital hand anomalies (syndactyly, cleft hand, etc.) had a shortened middle phalanx of the small finger only; however, some patients also had a shortened middle phalanx of the index finger. The patients with brachymesophalangia of two and five rays also had a tendency to have shortened middle phalanges of the index and small fingers. Some patients' anomalies, such as shortening of the middle phalanges in both the index and small fingers, did not adequately fit Bell's classification. The patients who had shortened metacarpals sometimes also had shortened middle and distal phalanges and a shortened metatarsus. Combinations of shortened phalanges and/or metacarpals were so numerous that it was impossible to fit them into any simple classification. (J HAND SURG 11A:829-36, 1986.)

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Shortening of the metacarpal and phalangeal bones (brachydactyly) may be found in various congenital hand anomalies, such as syndactyly, cleft hand, symbrachydactyly, and some malformation syndromes. However, an isolated brachymetacarpia or brachyphalangia is occasionally seen and brachymesophalangia 5 (shortening in the middle phalanx of the small finger) is frequently found in persons of Japanese origin.

When shortening is limited to one phalanx of one or two digits there is no functional disturbance and, mor-