Radial Longitudinal Deficiency

Epidemiology

- Results from a radial ray deficiency during embryological development.
- Incidence ranges from 1 in 30,000 to 1 in 100,000.
- Ranges from 4.7% to 6.1% of all congenital anomalies.
- Slightly more common in males than females and in caucasians.
- Bilateral in 38 to 50 percent of cases.
- When unilateral, occurs twice as frequent on right side.

Radial Clubhand

- Deformity is radial deviation of hand with a short forearm (50-75% the length of normal forearm).
- Almost always present at birth.
- Prominent knob at distal end of ulna.
- Thumb may be absent or severely deficient.
- Hand typically small.
- MCP joints with limited flexion and some hyperextensibility. May be flexion contractures of PIP joints.
- Elbow extension contracture common as a result of weak or absent elbow flexors.

Etiology

- Primary insult is to the apical ectodermal ridge during critical limb development period. (between 4^{th} and 7^{th} weeks)
- Mostly due to environmental factors.
  - Compression
  - Inflammatory processes
  - Nutritional deficiency
  - Irradiation
  - Infection
  - Chemical exposure
  - Medications (especially thalidomide)

Etiology

- No single definite implicating factors.
- Genetic studies have failed to show any genetic basis except when deformity is associated with a syndromal picture as in Holt-Oram, Fanconi's, and TAR syndrome (more later on these).
- Interestingly, one study found that twice as many affected patients born during summer months than winter months.

Bony Anatomy

- Scapula, clavicle, and humerus reduced in size.
- Ulna typically short, thick, and bowed. May develop synostosis with radial anlage.
- Scaphoid, trapezium absent in more than 50%.
- Lunate, trapezoid, and pisiform deficient in about 10%.
- Thumb metacarpal and its phalanges absent in more 80%, however a rudimentary thumb may be present.
- The capitate, hamate, triquetrum, and ulnar four metacarpals and phalanges present and without deficiency.

Muscle Anatomy

- Preaxial musculature from lateral epicondyle most severely affected.
- Radial wrist extensors (ECRL, ECRB and BR either absent or severely deficient.
- Finger extensors usually present.
- Typically see abnormal insertions of FCR.
- Long head of biceps almost always absent. Short head typically hypoplastic.
- Brachialis deficient or absent

Nerve Anatomy

- Median nerve thickened and runs just below fascia. At risk for injury during surgical dissection along concavity of deformity.
- Radial nerve typically ends at lateral epicondyle after innervating triceps.
- Ulnar nerve normal.
- MC nerve absent.

Vascular Anatomy

- Radial artery absent.
- Normal brachial and ulnar arteries.
- Well-developed common interosseous artery.

Associated Deformities

- Associated anomalies frequently encountered. These can be isolated or associated with a syndrome.
- These deformities include cardiac, GI, pulmonary, genitourinary, neurologic, and skeletal malformations.

Holt-Oram Syndrome

- Autosomal dominant mode of inheritance.
- See radial deficiencies of both thumb and radius.
- May see triphalangeal thumb, radioulnar synostosis, and proximal humeral abnormalities.
- The big association with this syndrome is cardiac defects.

Holt-Oram Syndrome

- Cardiac defects most frequently seen are ASD, VSD, tetralogy of Fallot, mitral valve prolapse, PDA, total anomalous pulmonary venous return.
- Congenital heart defects required for diagnosis.

TAR Syndrome

- Characterized by **T**hrombocytopenia and **A**bsent **R**adius.
- Thrombocytopenia begins in neonatal period.
- Thumb is **always present** and radial deficiency is **bilateral**.
- Autosomal recessive mode of inheritance.
- Typically, prognosis is good and platelet count improves to normal by age 4 to 5.
- Always check platelet count in child with Radial clubhand and a thumb prior to entertaining surgery.

Fanconi's Anemia

- Autosomal dominant
- Thumb always present
- A progressive pancytopenia
- May not progress until mid-childhood.
- Use mitomycin C test to diagnose or confirm.
- Prognosis is poor.

VATER

- **V**ertebral anomalies, **A**nal atresia, **T**racheo-esophageal fistula, **E**sophageal atresia, and **R**enal defects.
- Some or all of these abnormalities may present at the same time as radial deficiency.
- In some of these patients, a **C**ardiac anomaly, **L**ower limb defect, and **S**ingle umbilical artery is seen. (VACTERLS)

Trisomy 13 and 18

- Radial ray deficiencies seen frequently.
- However, prognosis is typically poor secondary to severe abnormalities.
- Surgical correction usually not indicated.

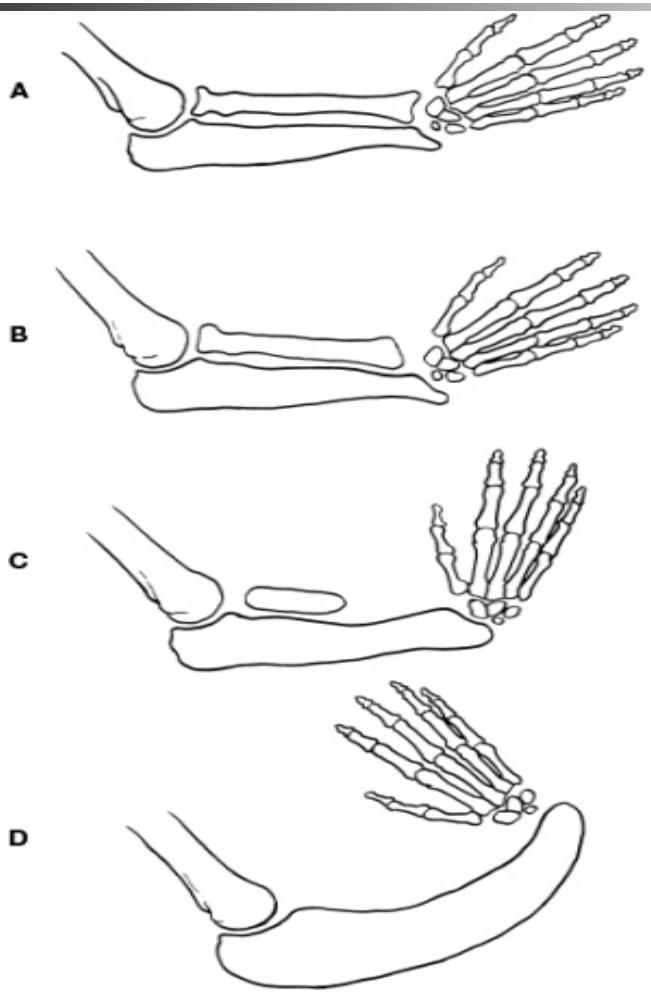
Diagnostic Considerations

- Absence or presence of thumb may help in differentiating syndrome from sporadic deficiency.
- Cardiac, hematologic, radiographic, and renal workup should be ordered on all children noted to have radial ray deficiency.

Classification

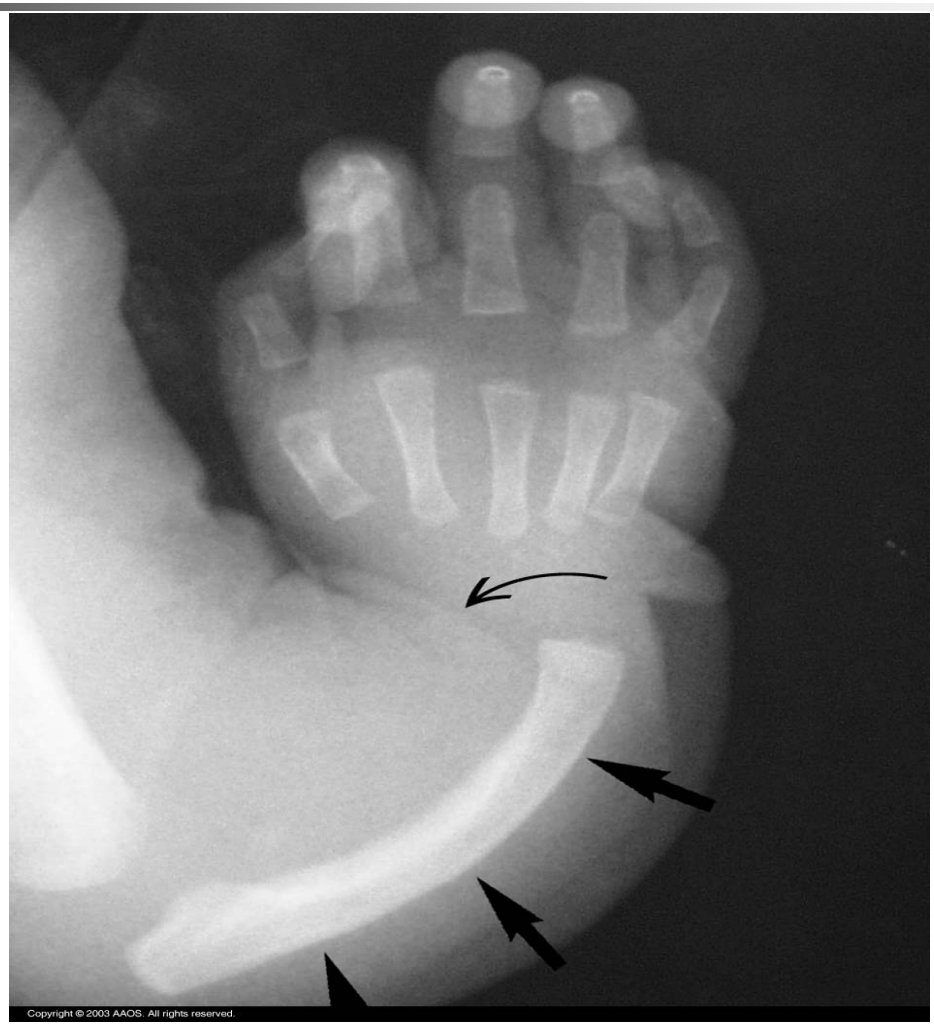
- Current classification described by Heikel.
  - Type I - Short distal radius. Distal physis present but delayed in appearance. Proximal physis normal.
  - Type II – Hypoplastic radius. Distal and proximal physes present but delayed in appearance.
  - Type III – Partial absence of radius. May be proximal, middle, or distal. (Distal 1/3 absence most common)
  - Type IV – Complete absence of radius

Classification



- Heikel's classification of radial dysplasia.
- **A,** Type I—short distal radius.
- **B,** Type II—hypoplastic radius.
- **C,** Type III—partial absence of radius.
- **D,** Type IV—total absence of radius

Xray



Non-Operative Treatment

- All treatment should begin at birth and should consist of manipulation and serial splinting/casting into corrected position.
- Should be done with a well-padded long arm cast with arm gently placed into flexion and hand in maximal correctable position.
- Cast on two weeks, then off with 1 week stretching/manipulation, then new cast x 2 weeks.

Non-Operative Treatment

- Riordan advocated a staged method of correction similar to the Ponseti technique for clubfoot.
  - Hand and wrist corrected first, then elbow corrected as much as possible.

Surgical Contraindications

- Life-threatening comorbidities
- Older patients with adequate function and psychological concerns.
- Bilateral involvement with stiff elbows.
- Type I and mild type II.

Pre-op Considerations

- It has been shown by Lamb, et al. that elbow flexion does not improve after surgical correction of hand.
- Be mindful that in patients with extension contracture at the elbow, correction of the hand will render them unable to reach their mouth.
- It is imperative that elbow motion be assessed and addressed prior to surgical correction.

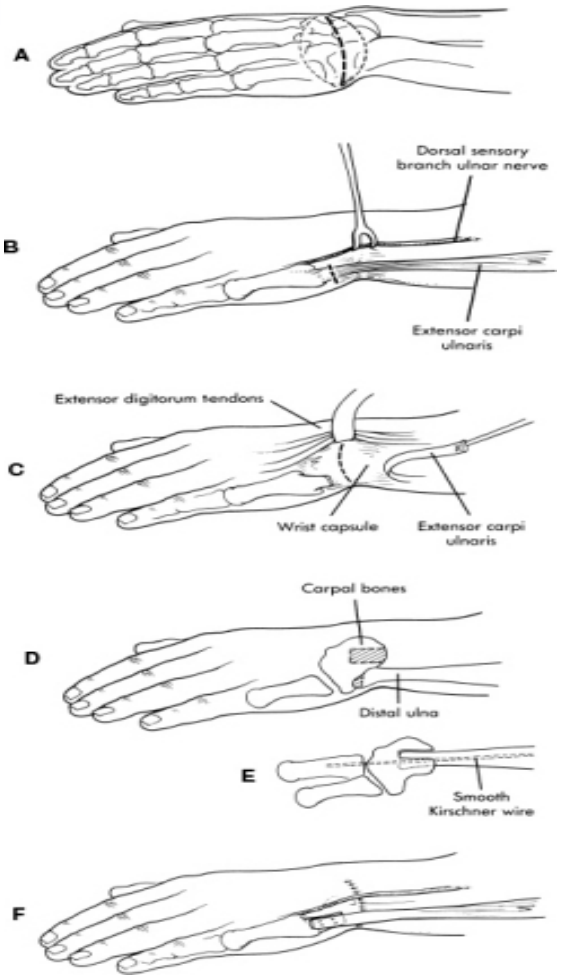
Operative Treatment

- In any case of operative treatment it is imperative that the carpus be able to mobilized over the end of the ulna.
- If this is not achievable after casting/splinting, lengthening through a distraction device or external fixator should be undertaken prior to formal fixation.

Operative Treatment

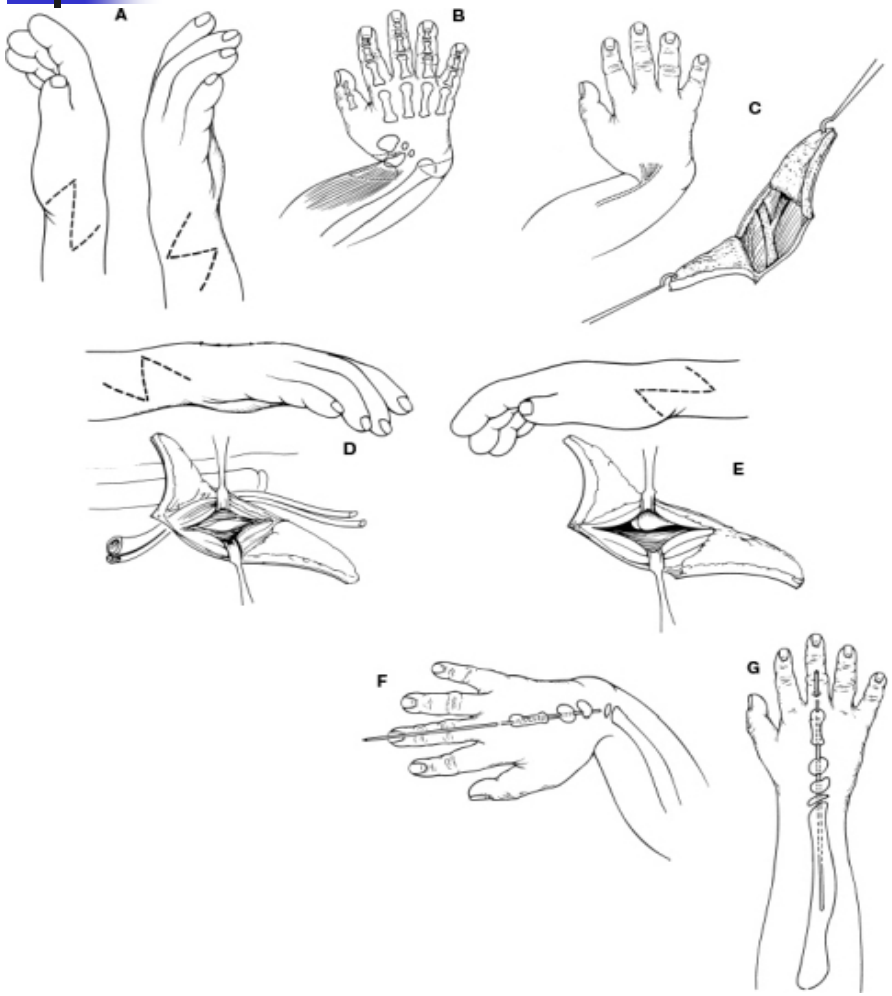
- Operative treatment largely involves the act of “centralizing” the carpus on the distal ulna.
- However, some feel centralization is not enough at may lead to recurrent deformity. These authors suggest “radialization” of the ulna.

Techniques



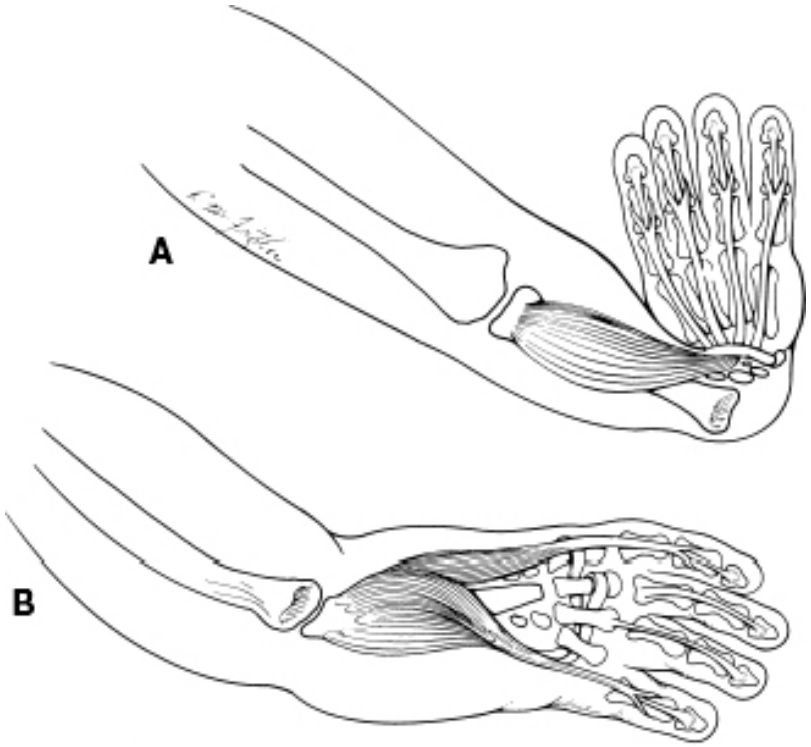
- Centralization arthroplasty technique, transverse ulnar approach.
- **A,** Incision.
- **B,** Exposure of muscle, tendon, and nerve.
- **C,** Capsular incision.
- **D,** Exposure of carpoulnar junction and excision of segment of carpal bones.
- **E,** Insertion of Kirschner wire.
- **F,** Reattachment of extensor carpi ulnaris tendon.

Techniques



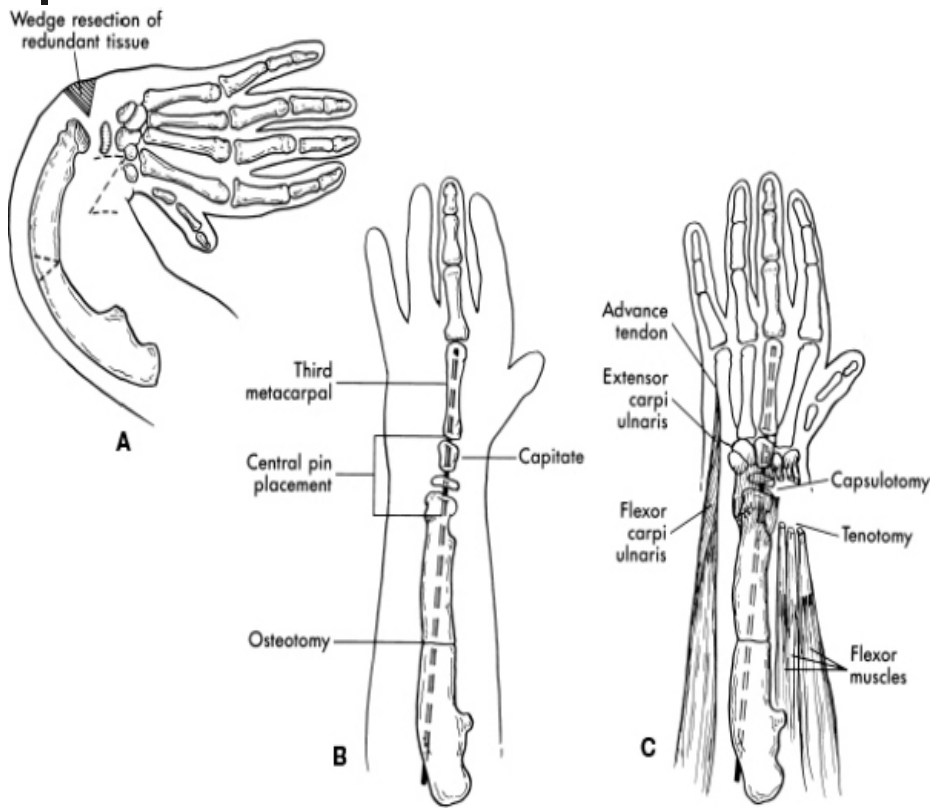
- Centralization of radial clubhand.
- A, Z-plasties on radial and ulnar sides of wrist.
- B, Incisions allow lengthening on radial side. Ulnar incision takes up skin redundancy, transposing it to deficient radial side.
- C, Radial incision in wrist for identification of median nerve.
- D, View from ulnar incision across wrist to radial incision after resection of all nonessential central structures.
- E, Distal ulna seen through radial incision at wrist.
- F, Kirschner wire passed through lunate, capitate, and long finger metacarpal.
- G, After centralization, Kirschner wire passed into ulna to maintain position

Techniques



- Centralization of hand and tendon transfer.
- **A,** Volar aspect of radial clubhand deformity showing right-angle relationship of hand and forearm and acute angulation of extrinsic flexor tendons.
- **B,** Volar aspect after centralization and transfer of sublimis tendons of ring and long fingers

Techniques



- Centralization of radial clubhand.
- **A,** Radial release and resection of redundant soft tissue.
- **B,** Centralization and pin fixation with ulnar osteotomy.
- **C,** Radial capsular release and tendon transfer.

Damore, et al.

- 19 radial club hands (16 type IV, 3 type III) treated with centralization followed over 6.5 years.
- “Average preoperative angulation measured 83° (range, 55° to 110°). Centralization corrected the angulation an average of 58° (range, 15° to 95°) to an average immediate postoperative total angulation of 25° (range, 5° to 60°). At the final follow-up examination there was a loss of 38° (range, 5° to 105°) and the total angulation increased to an average of 63° (range, 20° to 120°). The difference between the preoperative, postoperative, and follow-up angles was statistically significant.”

Techniques

- “Radialization” popularized by Buck-Gramcko in 1985.
- Improve the ulnar lever arm by creating a more ulnar deviation and transferring radial extensor and flexor tendons to the ulnar side of the carpus.
- This is thought to reverse imbalance of tight radial muscles and provide a better mechanical advantage.

Geck, et al.

- 29 limbs in 23 patients with an average follow-up period of 50 months treated with radialization or centralization and compared outcomes.
- Radialization was similar to modified centralization in the final outcome. Survivorship analysis was performed using revision as the end point, with a survivorship rate at 5 years of 67%.
- “These data offer support for the hypothesis that a more ulnar translation and an ulnar angulation of the wrist is a means of reducing the radial lever arm and thus the incidence of deformity recurrence and need for revision.”

Summary

- Always be mindful of associated syndromal patterns and concomitant medical problems when approaching radial clubhand deformity.
- Set reasonable treatment goals and counsel families on reasonable expectations.
- Know that elbow flexion typically does not improve after surgery, so it is imperative to address this pre-op in order to maintain ADLs.

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