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The treatment of longitudinal radial deficiency

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Abstract

Absence of the radius is the commonest of the longitudinal deficiencies but is itself quite an ususual congenital anomaly. No one surgeon is likely to encounter the condition more than once or twice in a career and there is a strong case for congenital upper limb conditions to be collected into special clinics.

The pathology and methods of surgical treatment are reviewed. It is a difficult condition to treat but there is no doubt that function and appearance can be improved (Pulvertaft, 1973). Pollicisation of the index finger should be considered and can often greatly improve the function.

Longitudinal deficiency

Radial Club Hand is the commonest of the major longitudinal deficiencies. It results from hypoplasia and complete or partial absence of the radius. Complete absence is the commonest and may be bilateral or unilateral. The condition was common in cases of thalidomide embryopathy when two-thirds of the radial deficiencies were bilateral. Its aetiology is otherwise unknown. It is usually sporadic but rarely may be associated with other congenital abnormalities: 1) Thrombocytopenia (the TAR syndrome). (It is the only time that absence of radius is associated with a near normal thumb); 2) Fanconi syndrome; 3) Atrial septal defect (the Holt-Oram syndrome). (This is the only condition where absence of the radius is hereditary); 4) VATER syndrome (vertebral defects, anal atersia, tracheo-oesophageal fistula, and radial and renal dysplasia).

The deformity is characterised by a radially deviated wrist and the forearm is short, only growing to between half and two-thirds of normal length (Fig. 1). The elbow usually lies extended at birth and there is a varying degree of reduced active and passive flexion at the elbow which often improves during the early years of life. Operation to correct the deformity should be avoided, particularly in bilateral cases, until the elbow flexes actively to at least 90 degrees.

The defects spread beyond the absence of the radius (Heikel, 1959) and stiffness of the elbow and there is stiffness and contracture in the metacarpophalangeal and proximal interphalangeal joints (Fig. 2). The scaphoid and trapezium often fail to develop and the first ray may be missing or represented by a simple "dangle thumb" (Pouce Flottant) (Fig. 3). This usually has a deficient metacarpal and no long tendons or thenar muscles. Growth potential is poor and reconstruction is not recommended. The radial forearm muscles are severely affected; the flexor pollicis longus is usually absent, the index profundus often absent or attenuated, though the flexor carpi radialis is usually present but the radial carpal extensors are poor or absent. The extensors to the index and middle fingers are usually hypoplastic and often have abnormal insertions limiting the metacarpophalangeal movement. Neuro-

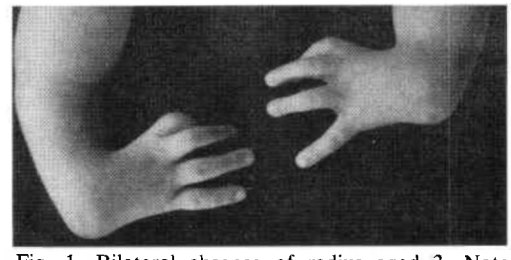

Fig. 1. Bilateral absence of radius aged 3. Note shortened forearm. The first ray of hand is absent and there is a four fingered hand with contracture of the radial two digits at the p.i.p. joints.

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Fig. 2. Radiograph showing absent radius and severe club hand deformity in the untreated wrist. No orthosis had been used to maintain the wrist straight and the deformity was uncorrectable.

vascular anomalies are frequent and the terminal part of the radial nerve may be missing and the area of sensation supplied by that nerve taken over by an abnormal division of the median. The radial vessels are often absent and also the radial digital artery to the index, which has implications if pollicisation is planned,

Treatment

The deformity is usually correctable at birth and gentle stretching by mother and therapist is indicated. Corrective splintage is difficult but once possible should be used at night and the baby allowed to use the hands normally during the day. These measures should be continued until a decision is made regarding operation. The wrist deformity is so ugly and the functional disability in bilateral cases so great that operation should be considered. The optimal time is between 6 and 9 months of age especially if it is intended later to pollicise the index finger.

Numerous operations described over the past 100 years have been inevitably followed by relapse of the deformity (Lamb, 1977). The operation with the best long-term improvement has been centralisation of the carpus (Lamb, 1977; Lidge 1969) (Fig. 4). Recurrence is liable during growth and an important part of the operation is transferring a major deforming force, by the flexor carpi radialis, to the dorsoulnar aspect of the wrist. Splintage of the wrist in the corrected position post-operatively is important and should be maintained for a minimum of 6 months and then at night for several years during growth.

Buck-Gramcko (1985) positions the ulna on the radial side of the carpus, (radialisation) thus avoiding resection of carpal bones.

Fig. 3. Dangle thumb (Pouce Flottant).

Fig. 4. Radiograph five years after centralisation of the carpus over lower ulna and four years after pollicisation of index finger.

Operation

The main steps are similar for both centralisation and radialisation. 1) An 'S' shaped incision is made from dorsum of hand to volar aspect of forearm. This gives good access to the carpus, the lower end of the ulna, the median nerve and the forearm muscles. 2) The extensor retinaculum is raised from radial to ulnar side and left attached to the ulnar side, and the extensor tendons exposed. 3) The median nerve is identified and any anomalous branch to the radial side. 4) The radial forearm muscles are exposed. The flexor carpi radialis can usually be identified but is sometimes joined with the radial carpal extensors. These muscles are removed from their insertion and will be transferred to the dorsi-ulnar aspect of the wrist later in the procedure. 5) An osteo-periosteal flap is raised from the dorsum of the carpus and the lower end of the ulna is dissected extra-periosteally without damage to the epiphyseal cartilage or its blood supply. Any fibrocartilaginous remnant of the radius (occasionally present in partial absence of radius) should be excised with all fibrous tissue preventing full passive repositioning of the hand over the ulna. 6) The next step depends on the nature of the procedure. If centralisation is intended the cartilaginous carpus is identified, sufficient of the central portions of this is removed to insert the lower ulna to a depth equal to its transverse diameter. This is in no way an arthrodesis. The intention is to preserve limited movement at this new ulno-carpal joint. The osteoperiosteal flap from the dorsum of the carpus is closed firmly and provides stability. It it is unstable or if any corrective osteotomy of the ulna is necessary, a Kirschner wire is inserted in a retrograde manner along the 3rd metacarpal shaft and then advanced across the centre of the lower end of the ulna and up the medullary cavity. This will keep the wrist correction and stabilise any ulnar osteotomy that was required. This wire can be left for several months but may extrude prematurely or break at wrist level with increasing activity. If radialisation is to be performed the ulna is positioned over the radial side of the carpus. A Kirschner wire is inserted obliquely through the second metacarpal and passed proximally along the shaft of the ulna. 7) The previously isolated radial muscles are transferred to the dorsi-ulnar aspect of the wrist and attached to the extensor carpi ulnaris. The retinaculum is repositioned and sutured in place.

Buck-Gramcko (1985) shortens the extensor carpi ulnaris by over-lapping and excises the redundant skin at the ulnar side. He reviewed 121 corrective procedures performed since 1969. Centralisation was carried out in the first 10 years and radialisation in the latter 10 years. Two-thirds had a subsequent pollicisation after the wrist operation. Results depend on the severity of the deformity, the condition of the muscles and function present in the fingers. He has little doubt that correction of the wrist is worthwhile, and that function is improved by pollicisation even though the quality of the index is seldom normal in this condition.

Lamb (1977) showed that children with bilateral deformity were unable to carry out all activities of daily living pre-operatively. Their overall function was significantly improved by correction of the wrist and pollicisation of the index finger.

Pollicisation (construction of a thumb)

A functional thumb is usually considered to contribute about 50 per cent of overall hand function. Attempts to reconstruct a functional thumb have therefore attracted much surgical endeavour (Littler, 1976). This is usually required for the following congenital conditions; 1) the four fingered hand; 2) the five fingered hand; 3) the "dangle thumb" with or without associated absence of the radius.

The technique evolved is based on transfer of the radial digit on its neurovascular bundle (Littler, 1953), shortening of the 2nd metacarpal and associated transposition of its interossei to reconstruct a new abductor and adductor of the thumb, and rotation of the retained 2nd metacarpal head so as to prevent an ugly hyperextension deformity of the thumb (Buck-Gramcko, 1971). Provided the structure and movement of the joints of the index finger and the quality of the intrinsic muscles of the 2nd ray are good the appearance and function of the index finger transposed in this way to the thumb position have given outstanding results.

Those surgeons who do not have the experience or expertise to obtain such results should refer children to a specialist unit.

Technique of operation

The operation in congenital absence is totally

different in concept and technique from operation for traumatic loss. Reconstruction of the whole thumb including the basal joint is required and is achieved by retaining the 2nd metacarpal head to form the new trapezium. Because of the wide range of movement at the metacarpophalangeal joint of children a hyperextension deformity is liable to develop at the base of the new thumb unless the metacarpal head is rotated into hyperextension so as to tighten the volar capsule.

Enough of the 2nd metacarpal shaft is removed so that the tip of the new thumb will reach the proximal interphalangeal joint of the long finger. If too much metacarpal shaft is preserved the thumb will look unnaturally long and be more a finger in the thumb position.

To obtain a good pinch between thumb and long finger the 2nd ray is rotated on its long axis about 150 degrees. The thumb should be abducted about 40 degrees.

If the intrinsic muscles of the 2nd ray are poor it will be necessary to reinforce these by an opposition transfer of the superficialis of the ring finger. This is seldom necessary with normal intrinsics which are stripped subperiosteally from the metacarpal shaft before its resection with careful preservation of their blood and nerve supply. These muscles are reinserted into the extensor aponeurosis over the original proximal interphalangeal joint which will now become the metacarpophalangeal joint. The first dorsal interosseus will become an abductor pollicis brevis and the first volar interosseus an adductor. The transposed radial intrinsic develops like a thenar eminence and in optimal cases the transposed index shows excellent mobility with full opposition and radial abduction (Fig. 5).

The optimal time for pollicisation is between 9 and 12 months of age. Buck-Gramcko (1971) has shown in 400 pollicisations for congenital cases that the end results are better the earlier the operation is performed. The transferred muscles hypertrophy, the bones of the index finger grow and broaden so that the proximal phalanx becomes like a first metacarpal and the child does not have to learn any new pattern of prehension.

For a good result the operation must be carried out meticulously and the surgeon have experience of working with small hands.

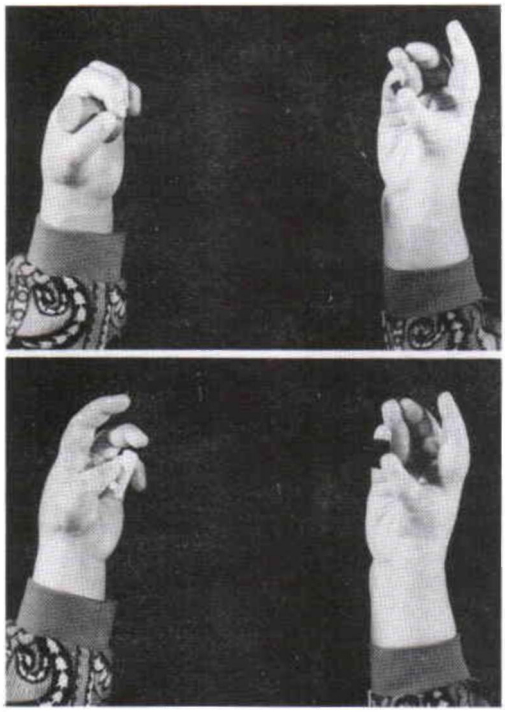

Fig. 5. Pollicisation of the left index finger at age 1 year. Excellent function with mobile thumb and good muscle control. Reconstruction right thumb with congenital absence thenar muscles.

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