We report the results of the treatment of 23 patients with macrodactyly. Eighteen had a two-stage bulk-reducing (defatting) procedure; phalangectomy was used to shorten the digits.

At a mean follow-up of nine years (2 to 12), two patients had been lost to follow-up, and three await a second-stage procedure. Good cosmetic correction was achieved in 12 patients, with satisfactory results in seven; two patients had poor results and required amputation.

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Macrodactyly is a rare congenital anomaly in which there is an increase in the size of all the elements of the digit, except the metacarpal or metatarsal. Barsky1 reviewed the 64 cases which had been described between 1840 and 1967. The reported incidence in the foot is low; most articles discuss only the hand.2

We report 23 patients treated for macrodactyly at our hospital between 1981 and 1996; in most the anomalies were in the foot (Fig. 1).

Patients and Methods

The series included 14 girls and nine boys seen at a mean age of five years (1 to 18). The fingers were affected in seven and the toes in 16. All the affected fingers were in the distribution of the median nerve (Fig. 2), but the toe anomalies were not associated with a particular nerve distribution. Enlargement of the digit was more pronounced distally and none of the patients had movement at interphalangeal joints. Three patients also had syndactyly (Fig. 3).

Table I gives the details of the various operations performed. Bulk reduction by defatting was done in two stages at an interval of three months. In the first stage, defatting was confined to one side of the digit, usually the convex, and reduced its thickness by about 10% to 20%. Care was taken not to remove too much fat from the dermis to preserve the vascularity of the overlying skin. At the second stage, defatting was performed on the other side of the digit with shortening of the bone. Most techniques of bone shortening remove only part of a phalanx,3 which does not effectively reduce the size of the digit. We developed a method of shortening in which an entire phalanx was removed (Fig. 4). After phalangectomy, the capsules of the adjacent joints were sutured and the digit stabilised by a Kirschner wire. The extensor tendon was shortened, but the remaining flexor tendon was left to adjust with time. The Kirschner wire was removed after four to six weeks.

Two patients had release of a syndactyly as a third-stage procedure after defatting. Two patients required a repeat operation for shortening.

We evaluated our results as: good, reduction of the digit by 50% or more; satisfactory, reduction by 25% to 50%, but with an angular deformity; and poor, cosmetically unacceptable requiring amputation.

Results

The mean follow-up was nine years (2 to 12). Since no patient had movement at interphalangeal joints our only aim was cosmetic correction. Eighteen of our 23 patients had two-stage defatting. Two patients were lost to follow-up after the first stage, and three are awaiting the second stage.

Of the 21 reviewed patients, the results were good in 12 (57.2%) (Figs 5 and 6), satisfactory in seven (33.3%) and poor in two (9.5%). Three patients developed an angular deformity of the digit due to the contraction of the scar on one side which required corrective osteotomy of the phalanx. Patients with good or satisfactory results were generally pleased with the operation, but two considered that the cosmetic correction was not acceptable and opted for amputation.

Skin-flap necrosis after defatting was seen in two patients and eventually healed after the application of dressings.
Discussion

Macrodactyly is a rare congenital anomaly, seen as 0.9% of all congenital anomalies of the upper limb, and reported to be more common in males. In our series, females predominated (61%). Over 300 cases of macrodactyly of fingers have been described, but only 60 involving toes have been reported. Associated syndactyly has been reported in 10% of cases, and was seen in three (13%) patients in our series.
Various operations have been described including carpal tunnel release, bulk reduction procedures, stripping of nerves, resection of nerves, epiphyseal arrest, finger shortening, and angulation osteotomy and amputation.

Macrodactyly involves an increase in the size of the phalanges, tendons, nerves, vessels, subcutaneous fat, finger nails and skin, but not of metacarpals or metatarsals. Treatment is not entirely satisfactory: the digits remain thick, heavy and somewhat ugly even after defatting. In our technique (Fig. 2) one whole phalanx is excised; this gives greater shortening of the digit. It has been reported that fingers stiffen even more with age, and we feel that the cosmetic improvement produced by phalangectomy is more important.

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References