CHAPTER **28**

SURGICAL INTERVENTION IN JUVENILE MACRODACTYLY

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INTRODUCTION

Pedal macrodactyly can be difficult to treat surgically and even once a surgical intervention has been attempted a few more surgeries are necessary. The goal of surgical intervention in macrodactyly is to achieve an esthetically and functionally-acceptable limb. The affected limb or digit is noted as being abnormally large within the first few years of life. Common difficulties include difficulty walking, inability to fit in normal shoe gear, which escalates if not treated to stiff toes, distal ulcers, and sometimes osteomyelitis. This condition is not only physically debilitating but also emotionally. It is not uncommon for adult patients with macrodactyly to opt for amputation versus salvage technique due to the years of discomfort and frustration.

HISTORY

Dr. Barsky's literature review from 1827-1967, was one of the earliest to shed some light on the rare condition of macrodactyly. This condition was classified as either primary or secondary. The primary macrodactyly involves enlargement of the skin, tendons, nerves, vessels, subcutaneus fat, nails, and phalanges, but not the metatarsal. Secondary macrodactyly however involves hypertrophy of the skin and soft tissue only.

A subcategorization of macrodactyly is based on comparing the growth rate of the affected toe versus the child's overall growth. Static macrodactyly includes an enlarged toe that is present at birth and increases in size proportionately with the child. Progressive macrodactyly involves an enlarged digit that grows faster than the child overall. The most common conditions found in association with this deformity include neurofibromatosis, Albright's dysplacia, lymphangioma, arteriovenous fistula, lipoma, hemangioma, Sturge-Weber syndrome, Proteus syndrome, and in utero disruption of growth limiting factors.

Surgical options focus on decreasing the size of the affected foot and allowing the patient to fit in normal shoes. Debulking or defatting is the most commonly-attempted procedure and as the name insinuates it involves bulk reduction of the involved soft tissue. Although a great procedure, used alone it is not sufficient for size reduction and only provides short-term success. It is only indicated in children and according to Kotwal et al in 1998, needs to be followed by another debulking procedure within 3 months and more often than not a phalangectomy. After a follow up of 9 years it was found that 57% of patients had good results, 33% satisfactory, and 9% poor results and opted for amputation.

Phalangectomy involves shortening of the affected toe and successfully decreases the length of the affected ray. This can be achieved via 3 very-well known methods initially described and perfected by Barsky, Tsuge, and Kotwall and Farooque. Barsky performed the phalangectomy by arthrodesis, Tsuge by distal phalanx removal with dorsal flap while Kotwal and Farooque by middle phalanx removal.

The metatarsal spread angle is a crucial indicator for affected ray resection. In the lesser rays of the foot a metatarsal spread angle of 10 or less is a good indicator for a debulking procedure and repeated diaphyseal shortening. In the severe case where the metatarsal spread angle is greater than 10, a ray resection is indicated. First Ray resection is never an option due to biomechanical importance during gait.

CASE PRESENTATION

The patient was a 6-year-old Hispanic female with an enlarged hallux on the left foot. The deformity was present since birth and grew proportionally with the patient. The patient's mother reported delayed ambulation and difficulty finding proper shoe gear. The patient had no osseous involvement and therefore underwent a debulking procedure.

CONCLUSION

Macrodactyly is a rare and debilitating condition that requires many surgical procedures that are indicated throughout different stages of the patient's lives. The literature however differs on approaches but agrees that debulking of the affected toe is one of the initial procedures that should be attempted. Debulking provides temporary relief and comfort for the patient while allowing them to have somewhat normal milestones development. This approach becomes more successful and more permanent later in life when used in conjunction with other osseous procedures.

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Figure 1. Preoperative view.

Figure 2. Marking the tissue flap.

Figure 3. Debulking the toe.

Figure 4. Removal of subcutaneous fat.

Figure 5. Adjustment of tendons.

Figure 6. Plantar incision and closure.

Figure 7. Dorsal incision and closure.