

Early Surgical Repair of Macroductyly

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We report a case of macroductyly of the foot in a 3-year-old girl. The standard treatment for this condition has been ablation. Studies of phalangeal resection, phalangeal amputation, syndactylization, digit shortening, tissue debulking, and osteotomy have shown varying results. In the case reported here, debulking combined with a shortening osteotomy was the treatment of choice. Although the pediatric patient may require additional surgeries because the deformity will continue to grow, early treatment has allowed this child the benefit of a functional, cosmetically appealing foot that can be fitted with normal footwear. (J Am Podiatr Med Assoc 94(5): 499-501, 2004)

Macroductyly is a rare condition of unknown origin that results in enlargement of one or more digits of the hands or feet. The anomaly occurs less frequently in the feet than in the hands.¹ DeValentine et al¹ reviewed the literature published through mid-1978 and found that 74 cases of macroductyly had been reported; 29 cases involved the lower extremities. A review of the world literature by Kalen et al² in 1988 reported 167 cases of macroductyly, with 52 involving the feet. The first podiatric medical case was reported by Annandale³ in 1866 when a 16-year-old girl presented with bilateral macroductyly of the great toes. The malformation was present at birth and increased in size as the child grew. He also noted that the arteries leading to the affected parts were very enlarged.

In 1967, Barsky⁴ reviewed the literature published between 1827 and 1967 in an effort to better understand the malformation. After this review of 140 years of literature, he determined that only 56 reports of

true, or primary, macroductyly existed. He added eight more cases to the literature. Barsky's definition of macroductyly, the standard used today, states that phalanges, tendons, nerves, vessels, subcutaneous fat, fingernails, and skin must all be enlarged. Hypertrophy of the skin and soft tissue alone does not constitute primary macroductyly and thus is referred to as secondary. This hypertrophy may be a result of conditions such as neurofibromatosis, Albright's dysplasia, lymphangioma, arteriovenous fistula, fibrous dysplasia, lipoma, hemangioma, and the Sturge-Weber and Proteus syndromes.⁵⁻⁷ Macroductyly has been further classified into two forms—static and progressive. The static form is noted at birth, and the size increases proportionately as the child grows. In the progressive form, the digit begins to grow at a faster rate than normal early childhood growth. An overgrowth of tissue of the digits and the appendage may also be noted.⁴

The etiology of macroductyly is elusive. Several theories have been proposed for the manifestation, including lipomatous degeneration, neurofibromatosis, nerve dysfunction, and *in utero* disruption of growth-limiting factors.^{4, 6} Pathologic evaluation of tissue samples has found that there is an unusual increase in nerve size and infiltration with fatty tissue. This fatty tissue, when found in a child, resembles

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that of adult subcutaneous fat.⁴ Radiographically, there will be advanced bony development, overgrowth of fatty tissue, and larger-than-normal digital arteries.^{6, 8} Clinically, in addition to enlargement of the digit, the skin is thicker and the joints may be stiff.¹

Treatment options have evolved to salvage the affected digits. The early literature recommended ablation, thus solving the immediate problem but creating biomechanical and psychological problems for the patient.⁷ Recently described treatment options include epiphyseal plate arrest, debulking, osteotomy, reduction syndactyly, and epiphysiodesis.^{9, 10} In our case, we chose to perform debulking and osteotomy procedures.

Case Report

A healthy 3-year-old girl presented with a congenital hypertrophic fourth toe with flexion contracture. The digit, which was noticeably enlarged at birth, grew proportionately with the child, who experienced normal development. There was no familial history of macrodactyly or neurofibromatosis. On physical examination, the patient was found to be healthy, with no evidence of hypertrophy other than in the fourth toe. Findings from the neurovascular examination were normal. The toe was found to be much longer and wider than the others, with flexion contracture (Fig. 1). Radiographs revealed abnormal bone anatomy and a very large soft-tissue shadow (Fig. 2). Surgical correction was recommended to make the foot more functional, more accommodating of footwear, and more cosmetically appealing.

Initially, the toe was debulked by means of an elliptic incision over the midline of the toe, and a large



Figure 1. Hypertrophy of the fourth digit of the right foot.



Figure 2. Anteroposterior radiograph showing abnormal bony anatomy and soft-tissue shadowing.

piece of full-thickness skin and abundant subcutaneous tissue were removed. Further debulking was performed as needed. The extensor tendon was split at the midline over the proximal phalanx, and approximately two-thirds of the proximal phalanx was removed (Fig. 3). Care was taken to avoid the growth plate and the articular surface. A Kirschner wire was placed from distal to proximal to hold the osteotomy reduced (Fig. 4). The skin was fashioned as needed to obtain appropriate closure. The patient was then placed in a short-leg walking cast. The pin was removed at 3-week follow-up, and the foot was casted for an additional 3 weeks. The child is now wearing normal footwear (Fig. 5).

Discussion

In this case, a 3-year-old girl presented with macrodactyly of the fourth toe and underwent successful debulking and osteotomy procedures. As most reported cases have involved patients in their teens and early adulthood, it is important to note that the condition is repairable in the pediatric patient as well. However, the procedures may be of temporary benefit, as the hypertrophy cannot be halted. Epiphysiodesis will probably be necessary when the child reaches 7 to 8 years of age. Sobel et al¹⁰ documented the case of a young girl with gigantism of the first and second digits who underwent debulking and osteotomy procedures at ages 8, 19, and 23 years.



Figure 3. Postoperative radiograph showing resection of the proximal phalanx and debulking of soft tissue.



Figure 5. At 6-week follow-up, the toe shows good signs of healing and the ability to be placed in normal footwear.

Simple destruction of the affected physis with a curette, followed by pinning with a Kirschner wire, should allow for correction of the length over time. A simple rule is to perform a panepiphysiodesis when the involved toe reaches the length of the toe of the same-sex parent. Simultaneous osteotomy may be used. To prevent neurovascular damage, approaching one side of the toe at a time, plantar or dorsal, in 3- to 6-month intervals is recommended.⁷ Toe or ray amputation should be reserved for patients with grotesque enlargement of the extremity after surgical

options are exhausted. Patients must be informed that although the procedures will allow for a functional foot that can be fitted with normal footwear, perfect cosmesis may not be obtained.

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Figure 4. Lateral radiograph showing Kirschner wire placement.