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"An Educational Asset of IFSSH"

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MACRODACTYLY: EVALUATION AND MANAGEMENT OF DIGITAL GIGANTISM

Hand Surgery Resource from the IFSSH continues to provide educational content focused on the evaluation and management of congenital hand differences. This newsletter highlights key principles in the assessment and treatment of macrodactyly.

Introduction

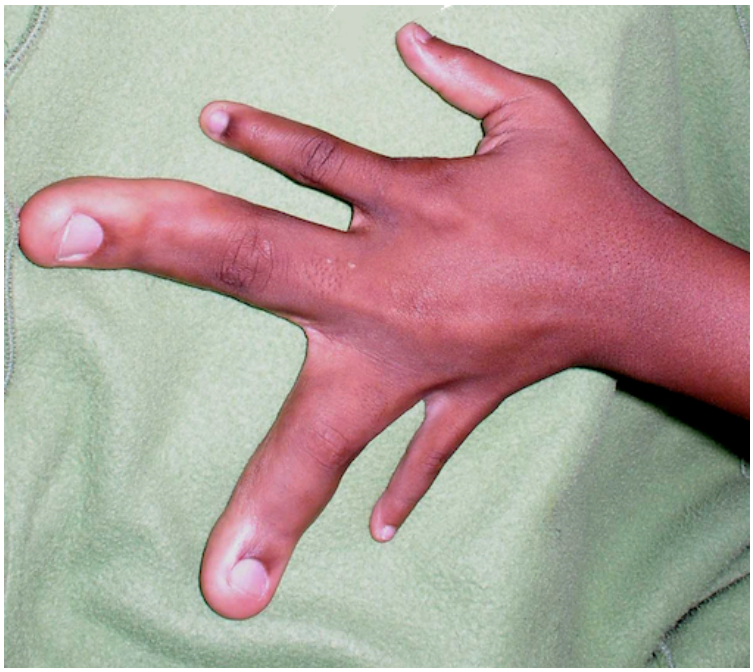
Macrodactyly, or digital gigantism, is a rare, nonhereditary congenital deformity marked by hyperplasia of one or more digits. In true cases, every structure of the affected digit is enlarged. Moderate to severe disease often causes pain, reduced ROM, and impaired hand function.

Pathophysiology

Proposed mechanisms include a neurogenic disorder (part of neurofibromatosis), loss of growth-limiting factors in development, tumorous overgrowth of one tissue (hemangioma, lymphangioma, enchondroma), and lipomatous degeneration. True macrodactyly (hamartomatous enlargement of all mesenchymal elements) is split into static (proportional growth from birth) and progressive (disproportional growth) types.

Related Anatomy

the long, thumb, ring, and little fingers. Adjoining metacarpals are typically spared.



Incidence and Related Conditions

Macrodactyly is about 0.9% of upper-extremity congenital anomalies and is more common in males (7:5). Associated conditions include Proteus, Bannayan-Riley-Ruvalcaba, Maffucci, and Klippel-Trenaunay-Weber syndromes, neurofibromatosis, lipofibromatous hamartoma of the median nerve, syndactyly, polydactyly, and clinodactyly.

Differential Diagnosis

Proteus syndrome, arteriovenous malformation, congenital lymphangioma, and macrodystrophia lipomatosa progressiva.

Symptoms

Enlarged digit(s), limited ROM, occasional pain, and reduced two-point discrimination. There is also significant potential for social and psychological problems.

Typical History

A typical case is a 16-year-old boy whose left long and long fingers grew faster than other digits, eventually dwarfing them. Progressive enlargement led to pain and trouble writing, driving, and opening doors, prompting surgery.

Examination

Workup Options

Plain x-rays show osseous overgrowth and joint status. MRI or angiography helps when a vascular malformation or nerve hamartoma is suspected. Prenatal ultrasound can detect it in utero.

Conservative Treatment

In mild cases without major enlargement or loss of function, observation alone is appropriate.

Operative Treatment

Severe or progressive disease often needs surgery. Age, type of overgrowth, digits involved, and patient and surgeon preference guide choices. Options include soft-tissue debulking, epiphysiodesis (most common), ostectomy, closing-wedge osteotomy, neurolysis, arthrodesis, and ray amputation for nonfunctional digits. Microvascular toe transfer may follow thumb amputation. Static must be distinguished from progressive disease early, as progressive cases need earlier surgery and more procedures.

Complications

Scarring, chronic pain, edema, reduced sensation, flexion contracture, hook nail, ungual deformity, IP joint stiffness, motion loss and recurrent enlargement of digits.

Outcomes

A truly normal finger is rarely achieved; counseling should emphasize that achieving an acceptable result not a perfect result most be the goal. Functional outcomes usually exceed aesthetic ones.

Key Educational Points

Congenital macrodactyly is the preferred term. It is one of the most difficult congenital hand anomalies to treat. Most children need more than one procedure, mainly for the progressive type. Recent research links somatic PIK3CA mutations to the PIK3CA-related overgrowth spectrum (PROS), and gene-targeted therapy is in development.

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