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## Updating Article

# Congenital deformities of the upper limbs. Part III: Overgrowth; Undergrowth; Streeter and others

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### A B S T R A C T

This article, presented in three sections, review the most commons upper limb malformations and their treatments. In this section three there's a discussion about overgrowth; undergrowth; Streeter Syndrome and other malformations. The bibliography is continuous since section one.

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## Introduction

This third and last article of this series, which had the objective of discussing the principles of treatments for congenital abnormalities of the upper limbs, covers the syndromes of overgrowth, undergrowth, Streeter and others that were not classified in the previous parts of this article.

### IV Overgrowth

This is exemplified by macrodactyly, which although rare still presents treatment challenges.<sup>44</sup> It represents a large number of distinct phenotypes with deformities both of bone and of soft tissue.<sup>44</sup> Its etiology remains unknown and therefore macrodactyly is evaluated within known syndromes, which include:<sup>44</sup>

- **Macrodactyly lipomatosa/macrodactyly within nerve tissues:** the commonest form. There is an accumulation of fatty tissue in the limb or finger affected. It is divided into a static type, when the overgrowth is proportional to the child's growth, and a progressive type, when this overgrowth is disproportional to the child's growth. The abnormalities affect tendons, intrinsic tissues, joints and phalanges.
- **Neurofibromatosis:** this is not a common cause. There are abnormalities in the peripheral nerves.
- **Proteus syndrome:** inexorable growth of the parts affected, with presence of exostosis, widening of the metacarpal heads, enchondromas, etc. There is no excessive fatty infiltration.
- **Hemihypertrophy/hemihyperplasia:** a genetic syndrome that is difficult to characterize. It generally affects one hemithorax or half of the body, and only rarely one finger in isolation.
- **Maffucci syndrome and Ollier disease:** multiple enchondromas with or without associated vascular malformations.
- **Klippel-Trénaunay:** vascular syndrome characterized by joint stiffness; it may be painful.

Because of the phenotypic differences between the various types of macrodactyly, there is no defined treatment algorithm. The treatment principles are that an esthetically acceptable hand with pincer capacity for gripping objects should be provided.<sup>44</sup>

Several techniques that combine osteotomy, epiphysiodesis, soft-tissue reduction (especially of fatty tissue), tendon shortening and neurectomy have been used with the aims of impeding overgrowth and reconstructing the appearance of the finger. Among these are the techniques described by Barsky, Tsuge, Hoshi, Milesi, Fujita, Ogino and Bertell.<sup>45,46</sup> However, none of these techniques, when applied to a large number of patients, has presented uniform long-term results.<sup>44</sup>

Amputation of the radius (Fig. 34) should still be considered as the treatment in cases of great disproportion of the segment affected and in cases of failure of the previous treatment.

Microsurgical transposition of a toe to the hand should be considered in cases of severe macrodactyly of the thumb or at the level of the metacarpus.<sup>44</sup>



**Fig. 34 - Macrodactyly. Resection of 3<sup>rd</sup> radius. Preoperative, intraoperative and final result.**

### V Undergrowth

The term brachydactyly means "short fingers". In cases in which brachydactyly is associated with syndactyly (union of two or more fingers), the term used is brachysyndactyly.<sup>47,48</sup> Other specific terms relating to shortening of the phalanges alone can also be found: brachyphalangism and ectrodactyly (absence of the phalanges); or shortening of the metacarpals: brachymetarpia or congenital short metacarpal.<sup>45,47,49</sup>

Cases that involve a phalange or metacarpal rarely need treatment. However, from an esthetic point of view, stretching can be performed with or without interposition of a bone graft, especially for the metacarpal.<sup>47</sup>

Blauth and Gekeler classified cases of brachysyndactyly as follows:<sup>47</sup>

- **Peromely:** transmetacarpal amputation.
- **Oligodactyly:** the central part of the hand is hypoplastic (hand with atypical cleft).
- **Short fingers:** all the fingers are short, with syndactyly, except for the thumb.
- **Monodactyly:** absence of the fingers; thumb present.

In many cases, there is no indication for surgical treatment because of good functioning.<sup>45</sup> In cases of poor hand function, the following can be indicated, depending on each case:<sup>47</sup>

- **Stretching of the metacarpals:** this is also an important tool for treating brachydactyly.
- **Transfer of phalange from the non-vascularized foot:** used in conjunction with local flaps as a compound graft. Greatly used, with several reports in the literature, but the results have been variable and inconsistent.
- **Microsurgical transfer of toe:** perhaps the best technique for recovering the pincer capability of the hand when only the thumb is present.

### VI Congenital constriction band syndrome (Streeter syndrome)

This is defined by intrauterine strangulation of parts of the fetus by the amniotic membrane, which causes deformities or even intrauterine amputation.<sup>50</sup> Its etiology remains undefined, but it may be due to vascular malformations that cause circumferential necrosis (replaced by fibrosis) in the limbs, or due to lesions in the amniotic membrane that adhere to and cause constriction of the limbs. The latter is the more accepted theory.<sup>45,50</sup>

Clinically, it is manifested by distal constrictions in the fingers, syndactyly, acrosyndactyly (with fenestrations), hypoplastic fingers and amputations.<sup>51</sup> Other malformations are found in 70% of the children with Streeter syndrome, clubfoot, lower-limb deformities, craniofacial defects, etc.<sup>50</sup> There may be neurovascular impairment and lesions in distal tendons. The central fingers are the ones most affected. The thumb is affected in only 21.5% of the cases.<sup>50</sup>

Some of the classifications that have been described are of little help in choosing the treatment. The classification most used is the one by Patterson:<sup>45,50</sup>

- 1) Presence of simple constriction bands.
- 2) Distal deformity at the bands, with or without lymphedema.
- 3) Bands accompanied by distal syndactyly (acrosyndactyly).
  - a. normal interdigital spaces.
  - b. shallow commissures; incomplete proximal syndactyly.
  - c. absence of commissures; small fenestrations separating the fingers.
- 4) Amputation.

Initially, Streeter syndrome may require emergency treatment, in cases of distal vascular distress at the constriction band, even if only a few hours or days after birth. Resection of the fibrous band (Fig. 35), below the subcutaneous level, is essential, followed by Z-plasty on the skin.<sup>50</sup>

Resection of the constriction bands can be done as a single surgical procedure (Fig. 36), provided that the venous return is preserved and that two constriction bands are not very close. This avoids distal congestion, which in more severe cases may lead to tissue necrosis.<sup>45,50</sup> This author prefers to release hemicircumferences in two distinct surgical procedures (Fig. 37).

In cases of complete syndactyly, the commissure needs to be reconstructed. Toe transfers, pollicization, finger stretching and “on-top plasty” need to be analyzed for each case, in amputations in which there may be a functional deficit.<sup>50</sup>

**VII Miscellaneous skeletal deformities**

**VII.1 Madelung**

Madelung deformity is defined as a growth deficit of the anteromedial portion of the growth plate of the distal third of the radius. In addition, there is shortening and curvature of the radius, widening of the distal radioulnar joint, dorsal subluxation of the ulna and triangular deformity of the carpus.<sup>46,52</sup> Some authors have believed that Madelung deformity is in fact a manifestation of dyschondrosteosis, and not an isolated malformation.<sup>46</sup>

The deformities are not noted until adolescence. They are generally bilateral and women are more affected than men. Its etiology remains uncertain, but vascular deficits and anomalous ligaments (Vickers) have been described as the cause of the deformity.<sup>46,52,53</sup>

No treatment is necessary for asymptomatic patients. For patients who complain of pain, orthoses and rehabilitation are often enough.<sup>53</sup>

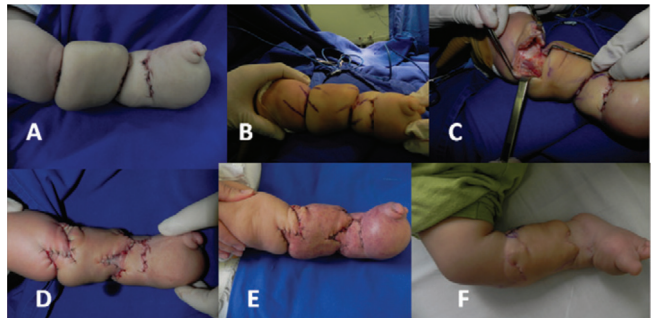
In cases of untreatable pain and limitations of range of motion that cause difficulties in activities of daily living, surgery is indicated. In the great majority of cases, the surgical indication comes from a static complaint.<sup>53</sup> During the growth phase, epiphysiolysis of the radius with interposition of fatty



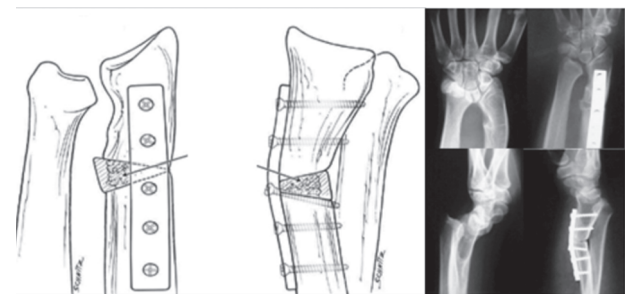
**Fig. 35 - Streeter syndrome with vascular impairment: preoperative, intraoperative, immediate postoperative and late postoperative periods.**



**Fig. 36 - Streeter syndrome: acrosyndactyly, before and after operation.**



**Fig. 37 - Streeter syndrome in a lower limb: before operation (A, B); during operation (C); after operation, with significant edema (D, E); and later on (F).**



**Fig. 38 - Technique for correction of Madelung deformity.<sup>52</sup>**

tissue and resection of the anomalous volar ligament (Vickers) has been described.<sup>46,53</sup> However, most authors have dealt with the deformity surgically in adulthood, after the growth plate has closed. Several techniques have been described: double osteotomy, single osteotomy, domed osteotomy, opening wedge, closing wedge, resection of head of the ulna and Sauvé-Kapandji surgery.<sup>46,52,53</sup> In our setting, de Paula et al.<sup>52</sup> described metadiaphyseal osteotomy with placement of a trapezoidal graft for correction of the deformity (Fig. 38).

## VII.2 Arthrogryposis

Arthrogryposis is a group of heterogenous syndromes that affect children and are characterized by congenital joint contractures in two or more joints. Between 104 and 300 syndromes have been described. The term amyoplasia describes classical arthrogryposis through correlating the joint contractures with the absence of intrauterine muscle function.<sup>54</sup>

The causes are multifactorial and include myopathy, neuropathy, connective tissue abnormalities, diminished intrauterine space, etc.<sup>51,54</sup> These conditions lead to a common denominator, which is the absence of intrauterine movement, which in turn causes the joint contractures and deformities.<sup>55</sup>

A distal form of arthrogryposis has been described, in which only the hands and feet are affected, thus sparing the major joints. Distal arthrogryposis responds well to conservative treatment consisting of stretching and serial orthoses, started soon after birth.<sup>54</sup>

Amyoplasia, which is the commonest form and is taken to be the classical form of arthrogryposis, has sporadic incidence and is characterized by symmetrical involvement of the joints, limbs of tubular appearance, absence of flexor skinfolds and normal intelligence. All four limbs are involved in 84% of the cases of amyoplasia, while only the lower limbs are involved in 11% and only the upper limbs in 5%.<sup>55</sup> Most of the patients present shoulders with internal rotation and adduction, extended elbows and wrists flexed with ulnar deviation. The fingers are flexed and rigid and the thumb is adducted.<sup>51,54</sup>

The upper limb should be dealt with as a functional unit, in the sense of promoting independence. The treatment priorities are to promote the capacity to communicate, perform activities of daily living, have mobility and be able to walk.<sup>51</sup>

The IFSSH (International Federation of Societies for Surgery of the Hand) recommends that manipulation, joint stretching and orthoses should be used soon after birth.<sup>51,54</sup> In relation to surgical treatment, there are some controversial studies in the literature in which the approach is made from the time when walking starts or after the age of 18 months. There are some authors who have recommended surgical interventions starting at the age of three months.<sup>51,54</sup>

Rotational osteotomy of the humerus may be indicated in some cases. The treatment for the wrist can be done using the following:

- Proximal carpectomy, when performed before ossification of the carpal bones, preserves the range of motion of the wrist through bone remodeling. In older children, recurrence is common.

- External fixator.
- V-shaped dorsal osteotomy: this corrects both of the deformities and preserves the growth plate of the radius. It is done on the mediocarpal joint, in association with stretching of the flexor tendons.
- Arthrodesis: this is reserved for mature skeletons.

Regarding the fingers, cases of an adducted thumb should be treated with reconstruction of the first commissure, stretching of the long flexor of the thumb and tenotomy of the adductor and first dorsal interosseous muscle, possibly followed by opponensplasty (Fig. 39). Metacarpal-phalangeal arthrodesis of the thumb may be necessary in maturity.<sup>51,54</sup> Finger rigidity is difficult to treat.<sup>51,54</sup>

Regarding the elbow, absence of active flexion is a major problem. The first step is to differentiate the cases with presence of passive flexion from those with a rigid elbow.<sup>56</sup> Any rigidity contraindicates tendon transfers. Techniques for posterior capsulotomy and/or triceps stretching have been described, in which the principle is assisted passive flexion, through which the patient uses the support of a table or bench in order to flex the elbows.<sup>51,56</sup>

Active flexion is a major challenge. The literature presents conflicting results relating to biceps-triceps transfer, pectoralis major-biceps transfer or latissimus dorsi-biceps transfer. Regarding transfer of the triceps to the biceps, the discussion centers on the fact that in the great majority of cases, loss of the triceps promotes deformity in flexion, i.e. the inverse of the original (Fig. 40). Elbow rigidity and the need to use the triceps for walking (crutches and wheelchair) are contraindications



**Fig. 39 - Adducted thumb treated by stretching the long flexor of the thumb; tenotomy of the thumb adductor and Kite flap.**



**Fig. 40 - Triceps-biceps after the operation. Limbs relaxed (A); attempt to raise the non-operated limb (B) and elevation of operated limb (C).**

for triceps-biceps transfer.<sup>51,54,56</sup> Even with transfers of the latissimus dorsi and pectoralis major, there is a long-term risk of contractures in flexion. Authors who have advocated these techniques have believed that positioning one limb in flexion and the other in extension would improve performance in activities of daily living.<sup>51,56,57</sup> Flexorplasty (Steindler) is another option for gaining elbow flexion in patients without any other options. The risk is that wrist and finger flexion is worsened.<sup>51,54</sup>

Clearly, there is still no consensus regarding treatments for elbows presenting arthrogryposis, and each case should be assessed and discussed exhaustively with the parents.

#### REFERENCES

44. Carty MJ, Taghinia A, Upton J. Overgrowth conditions: a diagnostic and therapeutic conundrum. *Hand Clin.* 2009;25(2):229-45.
45. Kay SP, McCombe D, Kozin SH. Deformities of the hand and the fingers. In: Green DP, Pederson MD, Hotchkiss RN, Wolf SW, editors. *Green's operative hand surgery.* 5th ed. Philadelphia: Churchill Livingstone/Elsevier; 2005. p. 1381-444.
46. Watt AJ, Chung KC. Generalized skeletal abnormalities. *Hand Clin.* 2009;25(2):265-76.
47. Nguyen ML, Jones NF. Undergrowth: brachydactyly. *Hand Clin.* 2009;25(2):247-55.
48. Samson P, Mevio G. Symbrachydactyly. *Chir Main.* 2008;27(Suppl 1):129-35.
49. Mattar Junior R. Deformidades congênitas do membro superior. *Acta Ortop Bras.* 1995;3(2):77-92.
50. Kawamura K, Chung KC. Constriction band syndrome. *Hand Clin.* 2009;25(2):257-64.
51. Ty JM, James MA. Failure of differentiation: Part II (arthrogryposis, camptodactyly, clinodactyly, madelung deformity, trigger finger, and trigger thumb). *Hand Clin.* 2009;25(2):195-213.
52. De Paula EJ, Cho AB, Junior RM, Zumiotti AV. Madelung's deformity: treatment with radial osteotomy and insertion of a trapezoidal wedge. *J Hand Surg Am.* 2006;31(7):1206-13.
53. James MA, Bednar M. Deformities of the wrist and forearm. In: Green DP, Pederson MD, Hotchkiss RN, Wolf SW, editors. *Green's operative hand surgery.* 5th ed. Philadelphia: Elsevier Churchill Livingstone; 2005. p. 1469-79.
54. Bevan WP, Hall JG, Bamshad M, Staheli LT, Jaffe KM, Song K. Arthrogryposis multiplex congenital (amyoplasia): an orthopaedic perspective. *J Pediatr Orthop.* 2007;27(5):594-600.
55. Katarincic JA. Congenital disorders: hypoplastic thumb. In: Berger RA, Weiss AP, editors. *Hand surgery.* Philadelphia: Lippincott Williams & Williams; 2004. p. 1445-51.
56. Van Heest A, James MA, Lewica A, Anderson KA. Posterior elbow capsulotomy with triceps lengthening for treatment of elbow extension contracture in children with arthrogryposis. *J Bone Joint Surg Am.* 2008;90(7):1517-23.
57. Lahoti O, Bell MJ. Transfer of pectoralis major in arthrogryposis to restore elbow flexion: deteriorating results in the long term. *J Bone Joint Surg Br.* 2005;87(6):858-60.