FUNDAÇÃO SÃO PAULO

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PROVA: CIRURGIA DA MÃO

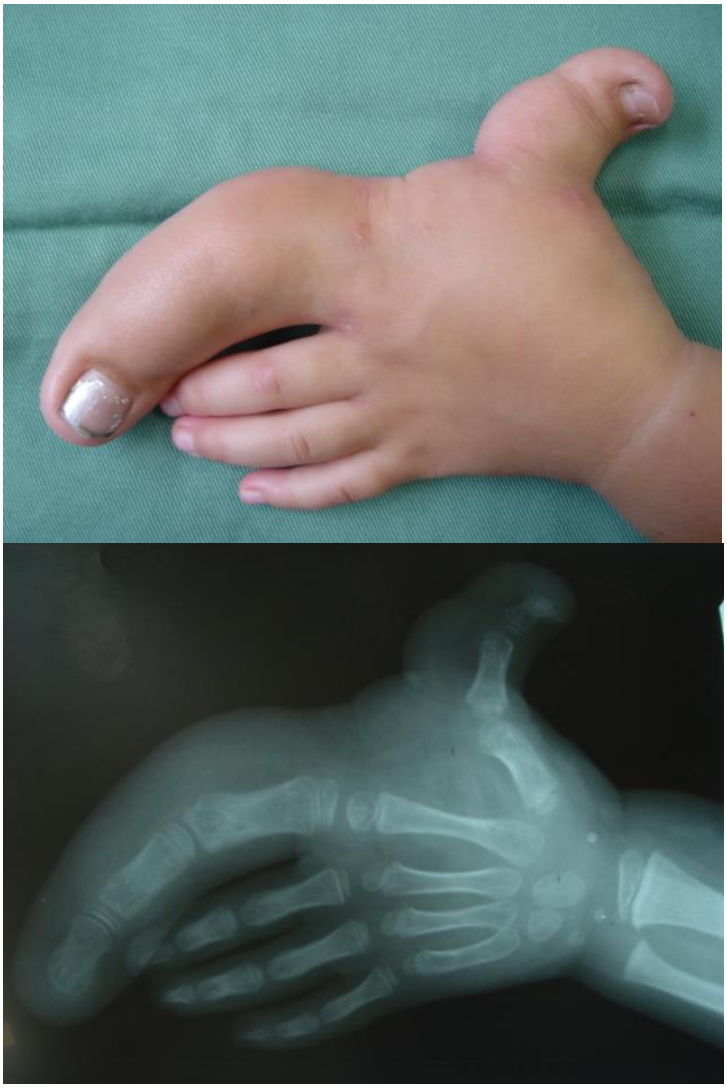
ESPECIALIDADE COM PRÉ-REQUISITO EM **ORTOPEDIA E TRAUMATOLOGIA OU CIRURGIA PLÁSTICA**

GABARITO DAS QUESTÕES OBJETIVAS

CIRURGIA DA MÃO	
1	A
2	D
3	B
4	C
5	B
6	C
7	A
8	D
9	A
10	C
11	D
12	D
13	C
14	B
15	C
16	C
17	A

CIRURGIA DA MÃO	
18	B
19	D
20	C
21	C
22	D
23	B
24	C
25	B
26	D
27	D
28	A
29	B
30	A
31	A
32	D
33	B
34	C
35	A

RESPOSTAS DAS QUESTÕES DISSERTATIVAS



A- Qual o diagnóstico?

Resposta – Macrodactilia

B- Classificações?

Resposta - Macrodactyly has been classified on the basis of rate of growth with Barsky proposing division of cases into a *static* type, where the enlarged digit grew commensurate with the growth of the child, and the *progressive* type, where the affected digit's growth outstrips the normal digital rate of growth.117 Flatt also proposed a classification based on the pathologic basis of the presentation distinguishing gigantism with lipofibromatosis (type I), gigantism with neurofibromatosis (type II), and gigantism with digital hyperostosis (type III).227 Upton later added

type IV, gigantism with hemihypertropy.233 The clinical usefulness of these systems is limited in practice by the variation in phenotype that is characteristic of the condition.234,235 There is value, however, in distinguishing the cases of overgrowth associated with specific syndromes because of the associated anomalies that may be expected and the importance of tumor surveillance in some of these syndromic patients.

C- Qual a etiologia e síndromes associadas a esta patologia?

Resposta - The advances in the understanding of the molecular biology of the condition are likely to illuminate the underlying pathology in macrodactyly and other forms of overgrowth. It is understood that macrodactyly forms part of the PIK3CA-related overgrowth spectrum (PROS) (see Box 36.2).236 Phosphatidylinositol 4,5-biphosphate 3-kinase Catalytic subunit alpha (PIK3CA) is the gene that codes for the production a component of the phosphatidylinositol 3-kinase (PI3K) protein that forms part of the PI3K-AKT- mTOR cell signaling pathway. Somatic mosaic gain-in- function mutations in PIK3CA in the developing limb bud can lead to uncontrolled growth of the affected part with the individual phenotype dependent on the timing and the tissue localization of the mutation.237 The PIK3CA mutation leading to macrodactyly leads to tumor-like growth, but it is a benign condition without potential for malignant transformation. The mutation is postzygotic and a sporadic condition without evidence of inheritance. Other overgrowth conditions in the PROS (see Box 36.2) have overlapping phenotypes with macrodactyly and have mutations in PIK3CA gene that can be identified with polymerase chain reaction (PCR) assay or genome sequencing in affected tissue.236 Future classifications are likely to be based on genomic analysis, with implications for natural history and perhaps nonsurgical management.238

Overgrouth syndromes
PIK3CA-related
overgrowth spectrum (PROS)
Macrodactyly
Hemihyperplasia multiple lipomatosis (HHML)
Congenital lipomatous overgrowth, vascular malformations, epidermal nevi, scoliosis/skeletal and spinal (CLOVES) syndrome
Fibroadipose infiltrating lipomatosis
Megalencephaly-capillary
malformation (MCAP)
Dysplastic megalencephaly (DMEG)
Proteus syndrome
PTEN hamartoma tumor syndrome (PHTS)
Type II segmental Cowden syndrome
Epidermal nevus syndrome
Neurofibromatosis type I (NF I)
Vascular malformations with overgrowth

Parkes-Weber
syndrome
Klippel-Trenaunay
syndrome
Maffucci syndrome

D- Quais as linhas gerais no tratamento cirúrgico desta entidade?

Resposta -

Goal of Treatment Procedure
Limiting growth Digital nerve stripping
Epiphysiodesis
Digit reduction Soft tissue Debulking
Skeletal Terminalization
Reposition nail unit on shortened skeleton
Palmar pedicle, (Barsky procedure)117
Dorsal pedicle, (Tsuge procedure)239
Nail island flap246
Resection of distal portion of nail and pulp Tsuge253
Hoshi254
Fujita255
Bertelli256
Correction of deviation Closing wedge osteotomy (combined with epiphysiodesis as required)
Thumb macrodactyly Metacarpophalangeal arthrodesis
Millesi procedure257
Amputation Ray amputation (with transposition of digit for central ray amputation)

E- Temos alguma opção terapêutica não cirúrgica atualmente?

Resposta - The advances in understanding the mechanism of overgrowth in macrodactyly with the PI3K-AKT-mTOR pathway of intracellular signaling implicated (see above) has revealed pharmaceutical options. Inhibitors of the upregulation of this pathway have been used in oncology, and there is prospect that similar inhibitors may limit overgrowth. A clinical study has been performed in a group of patients with congenital lipomatous (fatty) overgrowth, vascular malformations, epidermal nevi, and scoliosis/ skeletal/spinal anomalies (CLOVES) and PROS conditions treatedwith a specific PIK3CA inhibitor with encouraging results. The results are encouraging; however, many questions remain about Pharmaceutical therapy.252

Referência bibliográfica: Green's 8ª edição Part VI Pediatric Hand **CHAPTER 36** Deformities of the Hand and Fingers pags 1438 - 1453