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The patient's approach in congenital hand differences

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ABSTRACT

The patients with congenital hand differences need a special approach which is often complicated, as it involves the delicate handling of psychological issues, the crucial decision-making process concerning the treatment and its timing, as well as the careful prognosis of the evolution of the congenital difference with growth. Depending on the type of the congenital difference of the child, referral for genetic counseling is important, as is the meticulous clinical examination and consultation for the exclusion of concomitant anomalies of internal organs and life-threatening medical conditions. The physician has also to take in consideration the child's adaptation to its disability and the alterations that any correction might cause at functionality's expense, as well as the important issue of every function's cortical representation that changes with age.

KEY WORDS: Congenital hand differences, timing for surgery, radial club hand, Fanconi anemia, cortical representation

Introduction

The recently introduced word "differences" was established in the literature as a substitute to the terms "malformations" and "anomalies", in order to avoid the subsequent verbal discrimination of children born with abnormal hands and the psychological effects such abnormalities can cause. The professional approach of a young patient and his or her family in those cases is often complicated, as it involves the delicate handling of psychological issues, the important decision-making process concerning the treatment and its timing, as well as the careful prognosis of the evolution of the congenital difference with growth. The physician has to also take in consideration the child's adaptation to its disability and the alterations that any correction might cause at functionality's expense, as well as the important issue of every function's cortical representation that changes with age.

Parental responses to the child's hand abnormality

The birth of a child with a visible congenital difference influences its family in many ways. The parents usually go through phases of denial, anger and distress [1] in varying forms and intensity. These early responses reflect their sense of loss of an anticipated "perfect" baby. Additionally, a sense of lack of control and anxiety about future pregnancies is added to this loss. Another important matter is other people's opin-

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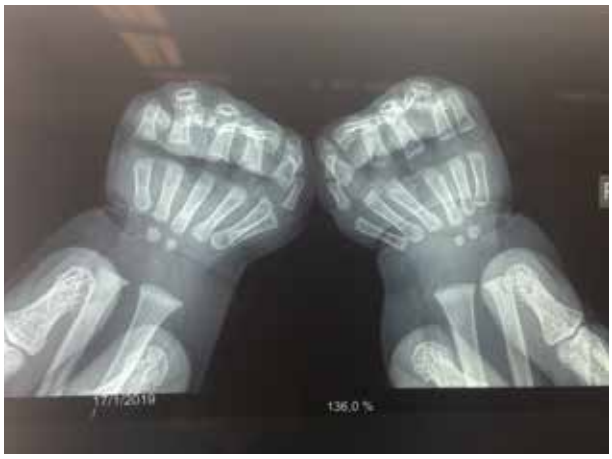

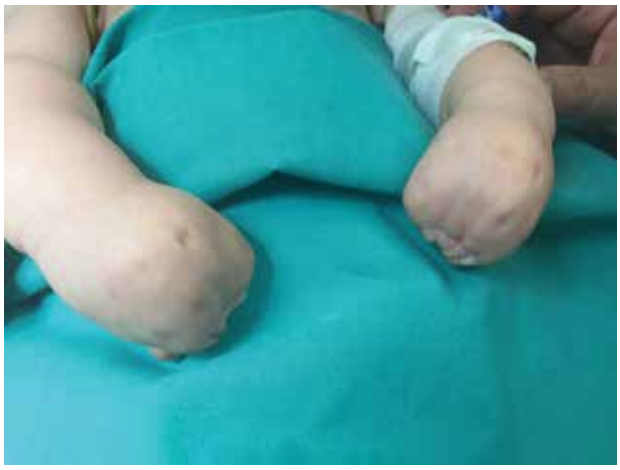

Picture 1: *Complex bilateral symbrachydactyly with polydactyly and early post-op results after four bilateral surgeries. The child's parents were informed from the beginning about the series of early surgical interventions needed and the results expected in terms of function and appearance.*

ions, including the response of family members to the child's malformation.

Although all parents feel enmeshed in the lives of their children and have a fundamental sense of responsibility for them, their course of adjustment to the difference of their baby's hand, varies [2]. Some are able to accept the situation and look for the best available treatment solutions, while others go through a long process of unresolved denial, grief and distress that emerges over the years. They usually feel a strong emotional sense of personal guilt and shame that leads to the tendency to hide the baby's hands. Furthermore, parents might encourage the child, as it grows, to keep the deformed hand covered, which has a devastating impact on the functionality of the whole upper extremity of the child due to lack of usage [8].

First consultation

During the first consultation, a thorough clinical examination must be conducted, followed by a detailed discussion with the parents. The best and most practical support that can be offered is a detailed and factual explanation of the incidence of these anomalies, their genetic heritage, the potential for growth, and the role of surgery in providing increased function and cosmetic improvement [3]. Depending on the type of the congenital difference of the child, the parents should be referred to genetic counseling and the young patient must be meticulously examined in order for concomitant anomalies of internal organs to be ruled out [9]. This is specifically important in cases of radial ray deficiency, which has been associated with almost every possible non-syndromic congenital abnormality as triphalangeal thumb, radio-ulnar synostosis, syndactyly, scoliosis, Sprengel deformity, club foot, congenital hip dislocation, ventricular septal defect, patent ductus anteriosus, lung abnormalities, cleft lip and palate, tracheoesophageal fistula, anal atresia, hydrocephalus, deafness, genitourinary anomalies and

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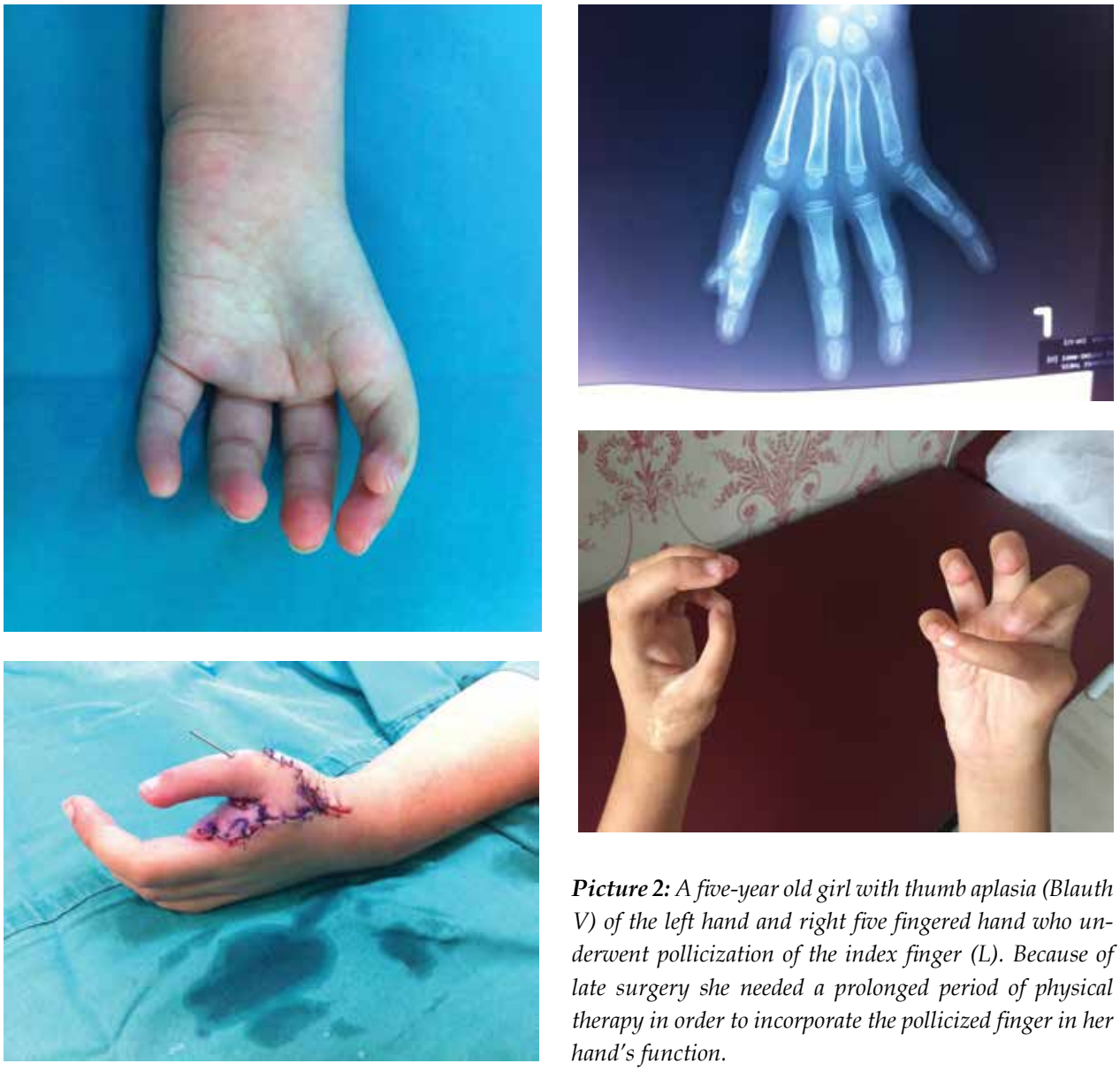

Picture 2: *A five-year old girl with thumb aplasia (Blauth V) of the left hand and right five fingered hand who underwent pollicization of the index finger (L). Because of late surgery she needed a prolonged period of physical therapy in order to incorporate the pollicized finger in her hand's function.*

other syndromes (TAR, Holt-Oram, VATER, Nager, Roberts and others). The most dangerous concomitant condition that needs attention is Fanconi anemia, which is an autosomal recessive disorder characterized by severe hypoplasia or aplasia of the bone marrow with anemia, thrombocytopenia and leukopenia. The pancytopenia usually develops progressively and appears between 5 and 10 years of age, with episodes of generalized purpura, bleeding and recurrent infections. Due to the fact that the prognosis is bad with survival only 2-5 years after the onset, and that bone marrow transplant is the only possible solution, the family and the attending pediatrician must both be alert, so that the child has frequent blood tests in order to achieve the earliest possible diagnosis. A hand surgeon's detailed information and counseling of a young patient's family about the danger of Fanconi anemia can actually save the child's life. In general indications for genetic counseling include: a known or suspected genetic disease in a patient or family, an individual who has a congenital malformation, unexplained mental retardation, advanced maternal age, abnormal prenatal screening tests, teratogen exposure and consanguinity. Genetic counseling should play a

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major role in the comprehensive medical management of a child with a hand malformation.

When discussing with the parents the treatment options, it is very important to define the goals of treatment and explain the specific methods that these goals can be achieved. Many children can clearly gain functional advantage from reconstructive surgery, but for some others the benefit is not always clear and the cost in terms of undergoing surgery and gaining secondary scars might be considerable. In cases that the anomaly's severity does not allow for reconstruction in a way that the hand would be more functional and cosmetically acceptable, the family should be informed about the cosmetic prostheses available, mainly for psychological reasons. The difficulty of balancing the needs of functional improvement with those of cosmesis should be emphasized and the fact that function is always prioritized over appearance should be made clear and be accepted. In case of different cultural priorities – for example cultures in which appearance is more important than function for the female hand – the parents should be given in detail the information that they need in order to make a decision themselves within that context. In addition, the optimal time for surgical intervention must be decided, bearing in mind all the developmental, social and psychological factors. The process of mutual understanding and shared decision-making involving the medical professional, the parents and the child, if appropriate, is absolutely necessary. The surgeon should respond to the needs, beliefs, and wishes of the parents, while also offering professional advice on the matter.

Timing for surgery

The three major goals of hand surgery are to enable the patient to orient his or her hand in space, to provide adequate and sensate skin cover, and to provide the patient a satisfactory power grasp with the ability to handle objects precisely [4]. A number of factors must be considered in organizing the timing of each procedure, including the psychosocial development of the child, the presence of other pathological conditions, the size of the structures to be operated on, and the normal growth and development of the hand [5]. Many congenital conditions as Apert syndrome, radial dysplasia and arthrogryposis require a complex planning of the reconstructions needed.

Historically, pediatricians have been reluctant to allow neonates to be subjected to surgery, due to their limited immune systems and the inherent risk of infection. Currently, urgent surgery is carried out in the first 5 weeks of life, while the child still has the passive immunity conferred by the mother, whereas less urgent procedures are delayed until the child is at least 5 months old, when active immunity has matured. Plans for surgical reconstruction should be designed to be completed by school age so that the child may adapt to and fully use the reconstructed limb instead of self-consciously trying to conceal it. [4]

Congenital differences treated in the immediate neonatal period

Simple procedures can be done conveniently in the neonatal period, such as ligating an extra digit attached by a skin tag or separating an acrosyndactyly connected by a minimal skin bridge. Severe constriction band syndrome that causes distal lymphedema and totally inhibits hand function should be treated by 10-12 weeks of age or ever earlier.

Congenital differences treated during the first year of life

The rationale for early surgery includes the avoidance of deformity and malfunction and the optimal use of infantile tissue plasticity. Syndactyly between border digits of the hand that have unequal length, syndactyly with bony bridges between terminal phalanges, acrosyndactyly with partial aplasia of the adjacent digit and club hand with partial or total radial aplasia are conditions that require early operation, possibly before the age of 6 months, to avoid a progressive bowing deformity. [4] Incongruent and irreversible joint changes may develop if certain conditions are not treated in a timely fashion, such as wedge-shaped triphalangeal thumb and congenital flexion contracture of the PIP joint. Early corrective surgery may occasionally be required for patients with camptodactyly, arthrogryposis and absent or deficient intrinsic musculature if no benefit has been obtained from splinting up to 6 months of age.

Early operation might be required not only because of the rapid growth that occurs in the first 2 years of life, but also because of functional consequences. Sur-

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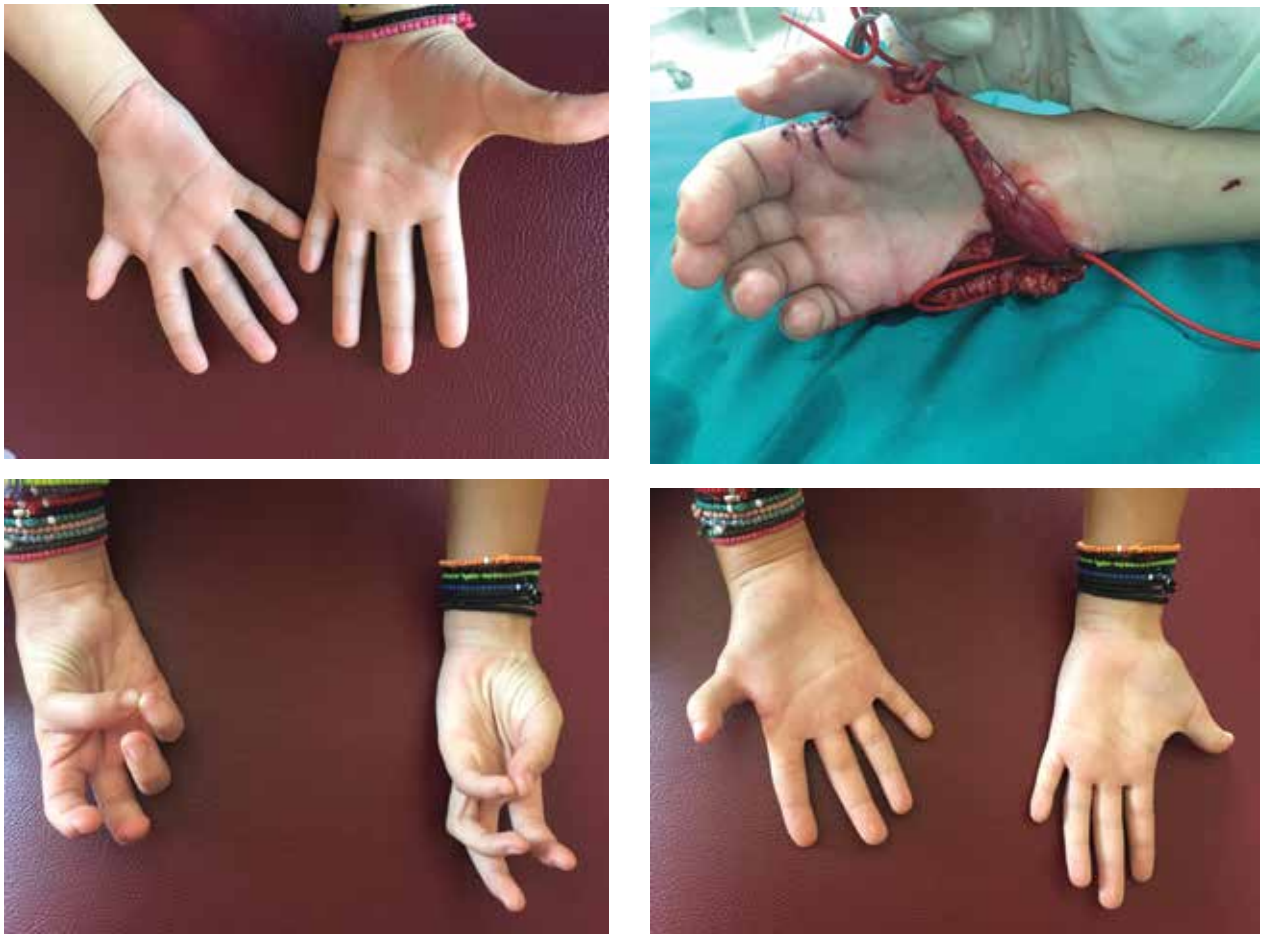

Picture 3: *A girl with Blauth II thumb hypoplasia that underwent Huber transfer with 1st web space Z-plasty at the age of 7 with excellent results.*

gery at a young age is considered mandatory in children with malformations in which hand function may be altered by surgery or in children at risk of developing certain grasping habits that would have to be altered by prolonged periods of physical therapy after corrective surgery, as in cases of cleft hand, proximal thumb duplication (Wassel IV) or pollicization of an index finger for aplasia or severe thumb hypoplasia. In all cases the physical ability of infant bones and soft tissues to adapt to changes produced by surgery in a key factor in deciding when to operate.

Congenital differences treated after the first year of life

The repair of an uncomplicated syndactyly, mild Blauth type II hypoplasia, symbrachydactyly and polydactyly can be put off until the second or third year of life, nevertheless surgery should be completed before school age, especially when several operations are required. One should not rush to treat conditions such as trigger thumb and minor clinodactyly because they may never require surgical correction. Other conditions that may be due to an underlying congenital anomaly with a strong genetic basis (such as certain types of camptodactyly) [7] manifest themselves only in older children. Complicated tendon transfers should generally be delayed until the child is 5-6 years old and able to cooperate in rehabilitation. The surgical treatment of some congenital hand deformities may have to be delayed beyond the first year of life because of life-threatening associated anomalies which must be treated and stabilized first.

Congenital differences that should not be treated

In certain conditions, surgical treatment may ac-

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tually be contraindicated. Lack of elbow flexion in a child with radial club hand is a contraindication to centralization of the hand, since the procedure would hamper the child's ability to move the hand to the mouth and would interfere with tasks as toileting. No treatment is indicated in unilateral or minor synostosis, since attempts to restore active rotation in radio-ulnar synostosis have been unsuccessful in the past. [6] In severe cases, a rotational osteotomy is done through the synostosis, at about 5 years of age.

The case of an older, well-adjusted child on whom surgical correction has not yet been undertaken, for whatever reason, should receive serious consideration, even in the presence of significant deformity, before the hand surgeon agrees to an operation. Soft tissues in older children do not adapt well, and functional patterns have already been established. An older child or a teenager should be psychologically evaluated and it must be explained that he or she should not have high expectations of achieving a normal hand.[7] Unrealistic prognoses are cruel, but because so much can be done for a child with a malformed hand, sympathetic optimism is completely justified. [3]

Conclusions

Surgeons specializing in the care of children's hand anomalies have the opportunity to treat not only the hand but also the child and the family. Understanding the psychological and social issues is an essential part of the care and treatment of the young patient. Early diagnosis of concurrent medical conditions or life threatening situations is of paramount importance. Timing of surgical interventions is also crucial and should be carefully determined. Children with congenital hand differences are challenging patients to care for, but they are undoubtedly the most rewarding, repaying success with years of function.

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