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Short Communication

Current Concepts in the Treatment of Ulnar Polydactyly

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Polydactyly is considered as one of the most common congenital hand anomalies affecting both the radial and the ulnar site.

Ulnar polydactyly has a greater incidence in patients of African heritage (1:143 live births) than in in Caucasians (1:1139). The real number of children suffering from this malformation may be underestimated, due to the fact that many patients, especially those affected by simple nubbins, are directly treated at birth with a ligation without consulting a hand surgeon [1].

Many cases are sporadic as consequence of a new mutation, even though, considering the fact that the association with other anomalies is frequent, genetic patterns may be found.

An autosomal dominant inheritance with incomplete penetrance and variable expressivity have been described, with numerous gene mutations on the chromosome 7, 13 and 19.

In case of syndromic association (Ellis Van Creveld syndrome or chondroectodermal dysplasia), especially in Caucasians, an autosomal recessive transmission has been reported [2].

Ulnar polydactyly has been for years considered in the group of duplications, inserted in Swanson's classification of congenital hand anomalies, commonly adopted by the International Federation of Society for Hand Surgery (IFSSH). However, a new classification system known as OMT classification, is now used: polydactyly, included in the group of malformations, arises from a failure of axis formation/differentiation of the hand plate in the radial/ulnar axis [3,4].

Among the different clinical forms, an undeveloped extradigit (nubbins) or a well-formed finger are the two most common ones.

In the past, many Authors tried of classifying the variable expression of the malformations.

In the sixties of the last century, Stelling categorized three possible forms, varying from a rudimentary nubbin to a well formed sixth finger.

In the late seventies, Temtamy and McKusick proposed an easy classification to differentiate the cases of complete duplication (type A) from those with rudimentary extra digits (type B).

More recently, Rayan and Frey, Al Quattan and Pritsch., *et al*. have published their own classifications.

Although their effort of classifying has been remarkable, the post-axial polydactyly presents the same problem of including in the classification all the clinical aspects, as it happens in preaxial forms.

With regard to this, in 2015 Duran., *et al*. described a new system with the purpose of including in a single classification all the aspects of these anomalies [5-10].

The examination should be focused on understanding the features of the polydactyly.

The range of motion of the fingers at CMC, MCP and IP levels should be observed during the natural movements of the hand. Moreover, a special attention has to be drawn in observing possible "en block" movements of the ulnar digits, resulting in stiffness of the fingers, particularly when cartilagineous connections have been arisen.

The examination of the hand should be extended to the entire upper limb in order to detect other anomalies of wrist, elbow and shoulder, especially in children with syndromic associations.

X-rays are mandatory to study the bone morphology and the joint alignment, even though some anomalies, as cartilagineous connections, cannot be well evaluated.

Some Authors recommend that surgery should be carried out before 6 and 9 months of age, before the acquisition of fine motor skills.

However, many surgeons prefer to postpone the beginning of the treatment because of the high risk of general anesthesia in the first 12 months of life. They suggest performing the operation between 12 and 18 months.

Whenever the duplication is due to simple nubbins, the removal can be carried out using a 4/0 not absorbable suture put at the base of the cutaneous bridge, as ligature. Alternatively, an elliptic incision has to be drawn at the base of the extra digit; initially the neurovascular bundle has to be isolated, then the nerve tractioned and transected and finally the artery coagulated [11].

Fully developed extra digits require a different approach as to recreate the stability and the motion of MCP joint. In fact, a simple amputation is destined to fail.

A zig zag skin incisions, starting from the CMC joint to the base of the ulnar finger, should be then performed. Some authors prefer to continue along the digit using the same incision, whereas the racket incision is the choice of others.

All the anatomical structures, namely skin flaps, extensor and flexor tendons, have to be isolated and elevated, exposing the MCP joint.

The distal insertion of hypothenar muscles, usually connected with the extradigit, needs to be preserved and elevated as well as the ulnar collateral ligament of MCP joint; however, a periosteum strip should be maintained and then re-attached.

The extradigit is now ready to be amputated allowing the exposition of the head of the V metacarpal bone, which is often bifid. At that level a chondroplasty, using a scalpel, is mandatory in order to obtain a better congruence of the articular surfaces.

Sometimes the neck of the metacarpal is angulated. In this event, a wedge closed osteotomy is the easiest way to achieve the right axis, setting the fragments with two Kirschner wires.

The ulnar collateral ligament and the hypothenar muscles have now to be reattached. A careful remodeling of skin flaps and sutures then carried out. This is the end of the operation.

A long arm cast will be maintained for 4 weeks when, after an X Ray control and the removal of Kirschner way, the active motion can start [12].

The outcome is generally satisfactory both for function and for cosmetic appearance. Additional surgical procedures are seldom requested.

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