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Review article

Management strategy for congenital thumb differences in paediatric patients

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ABSTRACT

Congenital thumb anomalies are common and have a major impact given the specific functional role of the thumb. They may occur alone or as part of a multiple congenital anomaly syndrome. The primary goal of surgical management is to improve or restore pincer grip. In patients with 'congenital' trigger thumb, the A1 pulley must be released if the interphalangeal joint remains in fixed flexion. Thumb duplication is generally managed by reconstruction of the thumb from the predominant (ulnar-based) digit; the accessory (radial-based) digit is excised after collection of its tissue components needed for the reconstruction programme. Thumb aplasia requires pollicisation of the index finger by island flap transfer of the second ray to give it the shape, position, and function of a thumb. Among patterns of digital hypoplasia, some require reconstruction of the existing thumb and others excision of the rudimentary thumb followed by pollicisation. In patients with aplasia of multiple hand digits, a toe transfer may be considered when there is no natural tendency to develop digital prehension at the hand.

1. Introduction

Congenital thumb anomalies are common and have a major impact given the crucial functional role of the thumb. When surgery is needed to profoundly change the prehension apparatus, the main procedure must be performed early, at about 12 months of age, to coincide with the development of the cerebral pathways that control grasp. Congenital thumb anomalies may occur in isolation (e.g., aplasia, hypoplasia, and duplication), in combination with other defects of the hand or upper limb (e.g., thumb hypoplasia with radial club hand), or as part of a multiple congenital anomaly syndrome (e.g., thumb aplasia in the VACTERL association). Any co-existing congenital anomalies must be detected before starting the surgical hand reconstruction programme. All the embryological mechanisms identified to date can be involved in the pathogenesis of thumb anomalies; we will specify the mechanism involved in the relevant section of this lecture. The primary objective of surgery is to improve or restore function. A source of considerable frustration after an ambitious and sophisticated thumb reconstruction programme is the child who, being left to his or her own devices, relies solely on lateral grasping between the fingers, neglecting the thumb that has been painstakingly rebuilt by the surgeon. Our objective here is to review the most common situations encountered by paediatric and hand surgeons, emphasising for each the main principles of management.

2. General paediatric assessment

When a patient presents with a congenital thumb anomaly, the surgeon must check that a general paediatric assessment has been performed.

The clinical assessment should include a thorough physical examination with a cardiac screen, inspection of the lower limbs (particularly the forefeet), and an evaluation of the hips.

First-line investigations consist of a comparative radiograph of the hands and affected thumb, echocardiography, renal ultrasonography, and a radiograph of the spine, in patients with clinical abnormalities or multiple birth defects.

Genetic counselling is offered routinely when the congenital anomaly does not exhibit the full range of characteristics indicating a generally sporadic event.

Multiple congenital anomaly syndromes that include anomalies of one or both thumbs are extremely numerous, and we will mention only the most common among them (Table 1).

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Table 1
Main multiple congenital anomaly syndromes that can involve the thumb.

Name of the Syndrome	Anomalies of the thumb	Other skeletal anomalies	Non-skeletal anomalies
Fanconi anaemia	Hypoplasia or aplasia	Radial club hand	Delayed development of haematological abnormalities
VACTERL association	Hypoplasia	Anomalies of the vertebrae (V) and limbs (L)	V: vertebral anomalies; A: anal atresia; C: cardiac defects; T: trachea-oesophageal fistula; E: Oesophageal atresia; R: renal and radial anomalies; and L: limb defects
Holt-Oram syndrome	Hypoplasia		Atrial septal defect
Goldenhar syndrome or Oculo-Auriculo-Vertebral (OAV) syndrome	Hypoplasia or aplasia	Vertebral anomalies	Dysmorphic facies, ocular and mandibular anomalies
Pfeiffer syndrome	Duplication	Duplication of the great toes	Anomaly of the first phalanx of the thumbs
Rubinstein-Taybi syndrome	Duplication	Duplication of the toes	Intellectual deficiency, abnormal facial features, microcephalus, glaucoma, calcaneal spurs
Greig syndrome or cephalo-poly-syndactyly syndrome	Duplication	Duplications of the toes	Scaphocephaly, frontal bossing, hypertelorism

3. Main clinical patterns

3.1. Trigger thumb

Trigger thumb, still often defined as congenital, is generally diagnosed in early childhood. This condition is ascribed to a growth mismatch between the flexor pollicis longus (FPL) and the A1 pulley that provides a fulcrum to the FPL tendon [1]. In most cases, the diagnosis is readily established based on fixed or mobile interphalangeal joint flexion with compensatory overextension of the metacarpo-phalangeal (MCP) joint and a palpable nodule (at the thickened A1 pulley) in the volar proximal digital flexion crease. Surgical release of the A1 pulley is appropriate in all patients who have a stable trigger thumb with no remission after several months.

3.2. Thumb duplication

Duplication is the second most common congenital thumb anomaly. In the classification developed by the American Society for Surgery of the Hand (ASSH) [2], thumb duplication is a Type III anomaly, i.e., an anomaly due to abnormal embryonic segmentation.

Recognising and analysing the duplication is the first step in the management strategy. The classification developed by Wassel [3] distinguishes several types based on the level of the duplication (Fig. 1). Although this classification is still in use, it is not sufficient to determine the principles of surgical management. Additional information required to that end is whether the duplication is symmetrical (with two digits of identical length and volume) or asymmetrical (with predominance of one digit, usually the ulnar-based digit) and whether there is malalignment in the coronal plane (clinodactyly) [4].

Simple excision of a small accessory digit is only very rarely performed. The main thumb is structurally normal, and its radial edge is attached by soft tissues to a floating thumb, which can easily be removed [5,6].

Choosing between midline fusion of the two digits (Bilhaut-Cloquet procedure [7]) and reconstruction based on one of the two digits is the next step in the management strategy. The choice is only theoretical, however, as midline fusion is now reserved for strictly symmetrical Type I, II, or III duplication, which is rare.

Reconstruction of a functioning thumb from one of the two digits is therefore the most widely used procedure and is performed in all cases of Type IV duplication, which is by far the most common variant (Fig. 2). In this complex technique, great care is given to a set of elementary procedures performed in combination with simple excision of the accessory (radial-based) digit. These procedures may require the use of tissue components taken from the radial-based digit before its excision [8]. They include the following four examples.

3.2.1. Augmentation

The ulnar-based digit, although larger than the radial-based digit, is smaller than the normal contralateral thumb. It can be augmented using tissue from the radial-based thumb collected before this last is excised. We advocate harvesting a long axial flap extending to the midline of the pad of the radial-based digit [9].

3.2.2. Correction of axial anomalies

Axial anomalies of the preserved ulnar-based digit are the rule. Angulation at the MCP joint is usually present. There may be another deviation, in the opposite direction, at the level of the interphalangeal joint. The deviation at the MCP joint is corrected by tensioning the ligaments that bridge the joint, whereas at the interphalangeal joint, an osteotomy, usually of the open-wedge type, is required. Any abnormalities in the distal attachments or course of the extrinsic flexors must be corrected to prevent recurrent clinodactyly.

3.2.3. Excision of bone and cartilage

The Wassel classification is based only on radiographic findings and therefore does not consider the details of the cartilaginous epiphysis division. These details are discovered intra-operatively, during arthrotomy of the MCP joint. They dictate the strategy for excising bone and cartilage. In most cases, bone and cartilage are removed from the radial edge of the metacarpal head, which is abnormally broad, as it initially articulates with the widened based of the duplicated first phalanx.

3.2.4. Re-attachment of muscles and ligaments

The collateral MCP ligament and the lateral thenar muscles are attached to the first phalanx of the radial-based digit, which is excised. To restore MCP stability, a flap should therefore be harvested from the radial-based digit before its resection. This composite flap includes periosteum, the collateral ligaments, and the attachments sites of the lateral thenar muscles. It should be inserted onto the main (ulnar-based) digit at the level of the first phalanx. Tensioning this flap corrects the deviation of the MCP joint space [10].

3.3. Thumb aplasia

The first step in the management consists in distinguishing between isolated thumb aplasia and thumb aplasia occurring as

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