Introduction

Congenital anomaly of the hand is an area that not every hand surgeon engages in routinely. It depends very much on the circumstantial situation whether the surgeon would have the experience to deal with the congenital problems. How do hand surgeons set priorities when dealing with congenital anomalies of the hand **(Hung et al, 2002).**

Congenital anomalies were often complicated structural abnormalities so that the question of priority would really deserve close scrutiny. Those who just performed the surgery according to what appeared convenient would not be able to provide the best results. On the contrary, an analytical prioritization before every surgical intervention would help significantly with functional restoration. Because we were dealing with children who presented with hand problems related to structures, there were obviously 3 areas of concern, namely, hand for surgery, children for surgery, and specific problems. The relationship between structure and function of the hand is so intimate that in any structural correction of the hand, the justification should be on a functional restoration, rather than a simple structural correction. In other words, with the exception of pure cosmetic corrections under very special circumstances, functional restoration or improvement overrules structural corrections. Functions of the hand should not be defined vaguely because they have long been identified and labeled. These functions include 3 different types of grip: power, diagonal, and hook; 3 different types of pinch: tip pinch, side pinch, and palp pinch, fulfilling different demands on physical activities, sensory feeling, and, lastly, expression. All surgical planning should pay full attention to the functional goal to be achieved. Although the physical ability of the individual has a lot to do with the functional achievement based on intact hand structure, one still has to observe that structural integrity is not everything. By that we are talking about the length and position of the digits, the mobility and stability of the joints, the strength of the components, and the sensibility of the tactile parts. Without a proper length and favorable position, no digit could function. Without stability and mobility, no joint could function. Without reasonable strength, no hand could function properly, and, without sensation, a hand would not be able to protect itself, not to mention precision performance **(Hung et al, 2002).**

These components need to be considered together. For example, good length without stability or mobility is meaningless.The importance of functional considerations in replantation surgery always has been a practical problem of concern. Although replanting severed parts of digits were technically straightforward and feasible under most circumstances, before engaging in the surgical procedure one has to seriously consider whether after the restoration of the structure the functional region was sufficient to justify the demanding procedures of replantation. Some recent reports on replantation surgery well supported the discussions. Cases of hand/finger replantation repeatedly were reported respectively: functional restorations were emphasized at the very beginning and details of actual functional return were given **(Bora et al, 1993)**.

All these practical relationships between structure and function should be considered during any surgical planning for the restoration of function in congenital anomalies .As far as surgical procedures are concerned, hand surgeons have worked out the order of priority to get the most ideal postoperative outcome. Different tissues deserve different amounts of attention. The order of priority should be: Vascular tissues for survival; Skin for rapid surface healing to allow other tissue healing and training; Bone for maintenance of basic functional structure,so that healing and training may be facilitated; Nerve for maintenance of sensibility; All other tissues do not received special priority attention because remedial measures are possible **(Razana & Hyzan**, **1998)**.

There are main principles governing pediatric management; first of all it would be wrong to consider children as small adults. The special considerations required for management of children arise from the fact that their bodies are still growing **(Lamb, 1972)**.

Performing structural correction as early as possible is a theoretical consideration that does not deserve serious support. The accepted principle should be as early as practical because too early is too difficult and might induce harm, whereas too late might mean too much growth damage and psychologic impairment. A good compromise is before 4 years unless severe growth disturbances do not allow the 3 to 4 years of deferral **(Lamb, 1972)**.

Congenital anomalies in the hand should not be considered as anomalies in vital organs that affect survival. Growth in the hand also is expected to be slower

than areas such as the maxillofacial area. Cleft lip and palate, for instance, would tend to shift open, if not repaired before 18 months. Hand anomalies, on the other hand, are not challenged by such early requirements. Moreover, the complexity of congenital anomalies also rules out the choice of having one major surgical endeavor to give the best chance of normal growth in the planning for correction **(Mark et al, 1991)**.

When the diagnosis is straightforward, surgical planning is simple. However, when the diagnosis is uncertain, planning will be very much affected. Thus,a hemangioma does not always require excision similar to other harmful tumors **(Mark et al, 1991)**.

Although old uncertainties and confused issues remained unsolved (e.g, over the different types of cleft hands), new forms of developmental syndromes with hand involvements are being reported. Japanese experts were particularly keen in the exposure of controversies existing in different forms of clefting. Cleft hands do have multiple varieties: missing longitudinal rays, total or partial; extra rays, complete or partial; abnormal joints and soft tissues; unstable to flail joints; and even hypoplasia.10 So in the overall diagnosis and planning of cleft hand corrections, comprehensive and thorough management requires a more academic and philosophic outlook to give the most reasonable outcome **(Benson et al, 1994)**.

Swanson tended to take a simplistic view that his classification of anomalies was still practical and comprehensive so that after a quick match of the pathologic condition, the most likely correct diagnosis could be made and then the management could be planned accordingly. The simple approach worked well for the simple situations in which a simple pathology was obvious. However, when multiple pathologies were present (eg, the coexistence of longitudinal defects, joint instability, rigidity, and tissue duplications), and when none of Swanson's clinical classifications fit well, clinical management became really difficult. Under such circumstances, identifying the most critical area and working out a staged policy would be a practical compromise **(Swanson & Swanson, 1983)**.

Carelessly performed surgery leads to epiphyseal damages, which leads to growth arrests and deformities. Correction of deformities needs to take into consideration the time dimension, not only the structural dimensions. Although growth plates and growing epiphysis were vulnerable to injury, it did not mean that

they could not be touched. In fact as long as the epiphyseal plate was untouched, the epiphysis could be cut flat and stitched to its adjacent component in an attempt to achieve linear fusion, trying not to affect the optimal growth **(Hung et al, 2002).**

Similarly, enlarged or abnormally faceted epiphyses could be shaved to give more congruent articulating surfaces without affecting the ultimate growth of the components handled. Tada and Yonenobi used this technique extensively in the Duplex thumb to bring about more satisfactory alignment of the retained component after sacrificing the unwanted component. This technique since the 1980s and have had very gratifying results **(Cheng & Leung, 1984)**.

Unlike an adult, young children do not tolerate pain and limitations of activities. Henceforth, dressing has to be double safe and better protected, pinning has to be secure and hidden, and casts need to go up one proximal joint (e.g, including the elbow) so as to avoid loosening **(Hung et al, 2002).**

It would be wrong to assume that severe psychologic disturbances exist in the child with severe anomalies whereas for those who are not so seriously affected (eg, duplication or syndactyly), psychologic considerations are not necessary. In fact, it depends on environmental conditions, individual circumstances, and immediate family support for the actual outcome of possible psychologic disturbances. It might be logical for every child suffering from a hand anomaly to receive a proper assessment from the psychologist **(Cohen & Kreiborg, 1995)**.

The varieties of imaging have expanded extensively in recent years so that congenital anomalies could acquire much benefits. Although the conventional radiography and angiogram still served routinely for structural identification and delineation of vascular pattern, disappointments were not uncommon. Ultrasonic examinations have benefited a lot of inflammation conditions such as tendonitis. The same device and technique should be useful to help identify thickened soft-tissue structures and to differentiate tissue planes between the normal and abnormal tissues. In one study, 75 surgical soft-tissue lesions of the hand were examined prospectively by experts on ultrasound and were correlated with surgical and pathologic findings. The normal ultrasonic anatomy of the hand was described. The use of real-time sonography allowed a reliable diagnosis of the cystic or solid nature of soft-tissue tumors, and accurate estimation of their volume and their precise 3-dimensional localization. Sonography facilitates the location of foreign

bodies, and appears as a new promising technique for the evaluation of tendons **(Schernberg et al, 1987)**.

Conventional tomography and computed tomography of the radioulnar joint and the wrist are used in patients with persisting complaints or doubtful findings on plain radiographs and difficult anatomic situations. Suspected ligamentous injuries of the wrist including tears of the triangular fibrocartilage complex are evaluated by wrist arthrography or magnetic resonance imaging, the latter requiring a highly skilled imaging and interpretation technique. Magnetic resonance imaging is the method of choice for the detection of osteonecrosis **(Al-qattan, 2001)**.

Aim of Work

The aim of this work was to discuss the diagnosis and the concepts in the management of congenital anomalies of the hand.

Embryology of the Upper Extremity

Embryogenesis

Limb development begins during embryogenesis with events that affect the position, number, and orientation of the limb. The limb bud is first visualized at 26 days after fertilization when the embryo is about 4 mm in length (crown-rump length), or about the size of a grain of rice **(Zaleske, 1985)**.

The bud rapidly develops through 47 days of life until the embryo is close to 20 mm in length, or the size of a lima bean. Fifty-two to 53 days after gestation, the embryo is between 22 and 24 mm in length, and the fingers are entirely separate . Eight weeks after fertilization, embryogenesis is complete and all limb structures are present **(Bamshad et al, 1999)**.

At this point, the joints develop by condensation of chondrogen to form dense plates between future bones. Joint cavitation further forms the articulation, although proper joint development requires motion for modeling of the ultimate joint surface. The majority of upper extremity congenital anomalies occur during this 4- to 8-week period of rapid and fragile limb development. After 8 weeks' gestation, the fetal period commences with differentiation, maturation, and enlargement of existing structures. The limb bud represents an outgrowth of the mesoderm into the overlying ectoderm. Two sources of cells migrate from their origins into the limb bud . The cells from the lateral plate mesoderm become bone, cartilage, and tendon. The cells from the somatic mesoderm form the muscle, nerve, and vascular elements of the limb bud (Figure 1) . To comprehend limb development and to explain it to families, the physician must appreciate the signaling centers that control the three spatial axes of limb development: proximal-distal, anterior-posterior, and dorsal-ventral **(Daluiski et al, 2001)**.

The signaling centers that control these different aspects of limb development are the apical ectodermal ridge (AER), the zone of polarizing activity (ZPA), and the Wnt (Wingless type) signaling centers (Table 1) **(Shubin et al, 1997)**.

A coordinated effort between the AER, ZPA, and Wnt pathways is necessary for proper limb patterning and axial development **(Niswander et al, 1994)**.

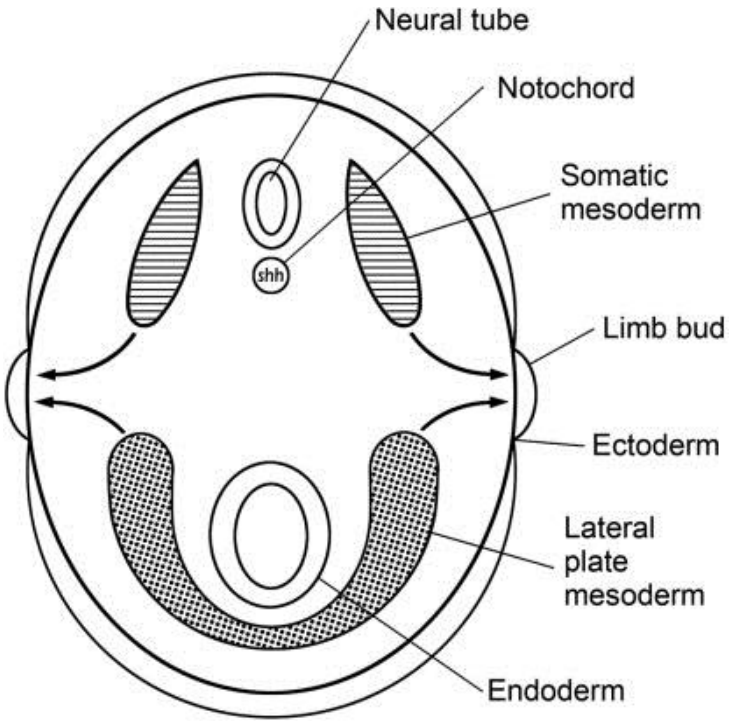

Figure 1: Axial view of an embryo. The lateral plate mesoderm forms the bone, cartilage, and tendon, and the somatic mesoderm forms the muscle, nerve, and vascular elements of the limb **(Riddle & Tabin, 1999)**.

The three signaling centers are interdependent such that loss of one signal results in compromise of the entire system **(Pearse & Tabin, 1998)**.

Signaling pathways during embryogenesis:

Signaling Center	Responsible Substance	Action	Anomaly
Apical ectodermal ridge	Fibroblast growth factors	Proximal to distal limb development, interdigital necrosis	Transverse deficiency
Zone of polarizing activity	Sonic hedgehog protein	Radioulnar limb formation	Mirror hand
Wnt pathway	Transcription factor, Lmx-1	Ventral and dorsal limb axis	Nail-patella syndrome, abnormal nail and pulp arrangement

Table 1: Signaling pathways during embryogenesis **(Shubin et al, 1997)**.

Programmed cell death (Apoptosis) plays an important role in limb development. It is an active process that is genetically controlled to eliminate unwanted cells during embryogenesis **(Chen & Zhao, 1998)**.

Apoptotic cells undergo a degenerative process consisting of DNA fragmentation and are eventually engulfed by phagocytic cells. Genetic control of cell death is necessary during limb bud formation. For example, interdigital necrosis is necessary for finger separation. Failure of interdigital apoptosis results in syndactyly **(Zakeri et al, 1994)**.

Interestingly, bone morphogenetic proteins (BMPs) are involved in many aspects of vertebrate development, and certain BMPs trigger apoptotic pathways **(Chen & Zhao, 1998)**.

Spatial axes of limb development and their signaling centers:

Proximal-Distal Limb Development

The limb develops in a proximal to distal direction, with the shoulder forming before the elbow and the elbow before the wrist. This progression is controlled by the AER, a thickened layer of ectoderm that condenses over the limb bud **(Daluiski et al, 2001)**.

This signaling center guides the underlying mesoderm to differentiate into appropriate structures **(Bamshad et al, 1999)**.

Removal of the AER results in limb truncation, and ectopic implantation of the AER produces additional limb formation (Figures 2 and 3) **(Tickle, 2004)**.

The secreted proteins within the AER that yield this effect are fibroblast growth factors **(Niswander & Martin, 1992)**.

In fact, removal of the AER can be overcome by the application of fibroblast growth factors . Mice deficient in various fibroblast growth factors have complete transverse limb defects **(Sekine et al, 1999)**.

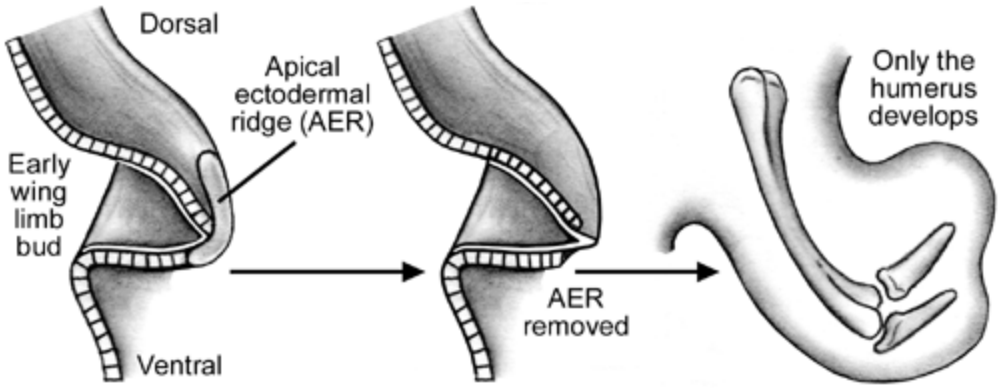

Figure 2: Removal of the AER from the developing limb bud results in limb truncation **(Riddle & Tabin , 1999)**.

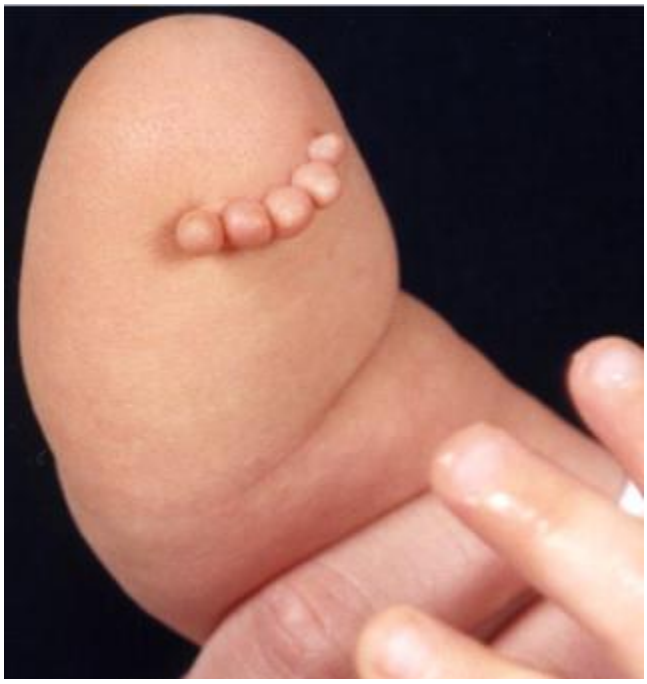

Figure 3: Short below-elbow transverse deficiency with residual nubbins attributed to loss of AER function **(Tickle, 2004)**.

When explaining this to a parent, we know that the AER was not physically removed. However, we do have evidence to suggest that bleeding or ischemia within the AER results in failure of the AER to work properly **(Gonzalez et al, 1998)**.

Transverse deficiencies are usually sporadic without any underlying reason. Concern for exposure to a teratogen is raised when multiple limbs are involved, a finding suggestive of a widespread insult to all developing limb buds. Transverse differences are not inheritable, and future children are unlikely to be affected.

Anterior-Posterior Limb Development

The limb also develops in an anterior-posterior (also called radioulnar or preaxial-postaxial) direction. The ZPA resides within the posterior margin of the limb bud and functions as a signaling center for anterior to posterior limb development **(Bamshad et al, 1999)**.

This signaling pathway polarizes the limb into a radial margin and an ulnar border **(Tickle, 1996)**.

The signaling molecule within this pathway is the sonic hedgehog compound.Transplantation of the ZPA or sonic hedgehog protein causes mirror duplication of the ulnar aspect of the limb (Figure 4) **(Daluiski et al, 2001)**.

The extent of duplication is dose dependent, with greater transference resulting in more replication . This explains the variable numbers of fingers and different phenotypes of mirror hands (Table 2) **(Al-Qattan et al, 1998)**.

Mutant mice that express sonic hedgehog protein in the anterior limb bud are polydactylous with duplication of their ulnar digits **(Masayu et al, 1995)**.

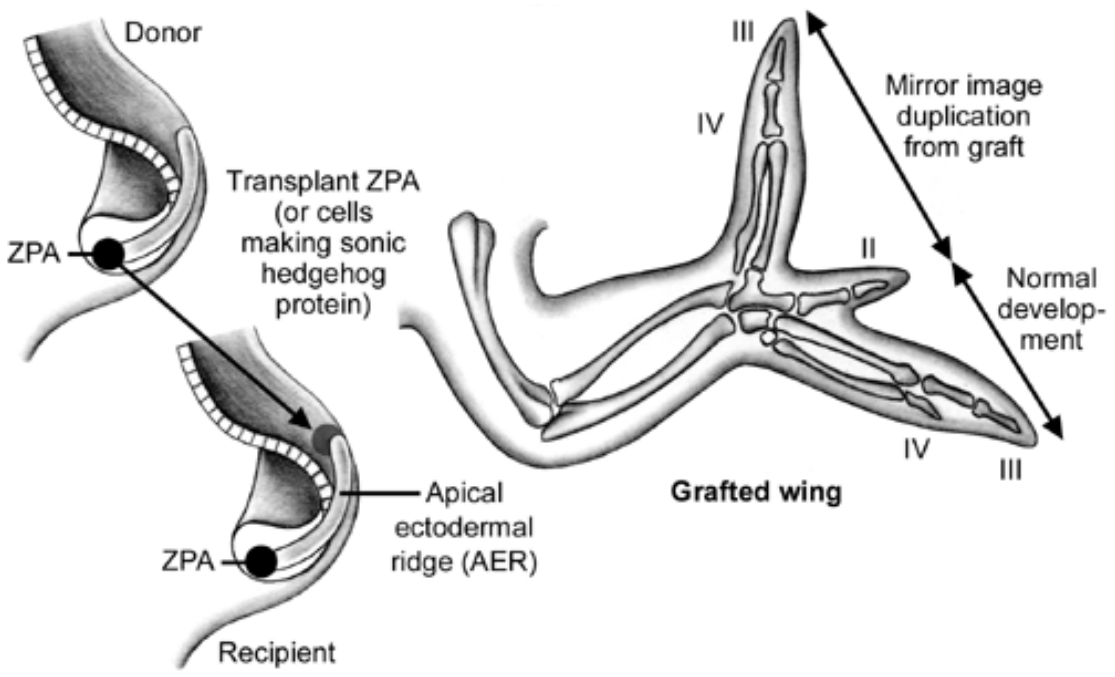

Figure 4: Transplantation of the ZPA or sonic hedgehog protein causes mirror image duplication of the ulnar aspect of the limb **(Riddle & Tabin , 1999)**.

Classification of Mirror Hands

Type	Name	Clinical Features
1	Ulnar dimelia	Multiple fingers with two ulnae Type A: Each ulna well formed Type B: Preaxial ulna hypoplastic
2	Intermediate form	Multiple fingers with two ulnae and a radius
3	Intermediate form	Multiple fingers with one ulna and a radius Type A: Radius well formed Type B: Hypoplastic radius
4	Syndromic form	Bilateral, mirror feet and nasal defects characteristic Type A: Sandrow's syndrome—two ulnae Type B: Martin's syndrome—an ulna and a radius
5	Multiple hand	Complete duplication of the hand, including the thumb; forearm normal

Table 2: Classification of mirror hands **(Al-Qattan et al, 1998)**.

Dorsal-ventral limb development

Dorsal-ventral limb development, or the process of differentiation between the dorsum of the finger with a fingernail and the volar surface abundant with pulp tissue, is not as well understood **(Daluiski et al, 2001)**.

The Wnt signaling pathway resides in the dorsal ectoderm and controls this spatial development. The pathway produces a transcription factor, Lmx-1, that induces the mesoderm to adopt dorsal characteristics **(Riddle et al, 1995)**.

In the ventral ectoderm, the Wnt pathway is blocked by a product of the gene Engrailed-1 (En-1). Mice lacking the Wnt pathway have ventralization of the dorsal surface (i.e., biventral limbs with palmar pads on both sides of the foot) **(Johnson & Tabin, 1997)**.

In contrast, in mice lacking En-1, dorsalization of the volar surface develops (also called bidorsal limbs) **(Loomis et al, 1996)**.

Differences affecting this pathway are relatively rare. Loss of Lmx-1 is associated with nail-patella syndrome. Occasionally, children will have anomalies consisting of an extraneous nail or abnormal pulp development. These abnormalities are related to the Wnt signaling pathway and occur sporadically without any definable cause **(Dreyer et al, 1998)**.

Anatomy

Integument/Fascia:

The palmar and dorsal skins are distinctively different with respect to structure and function. The palmar skin is thick with fibrous septa connecting the skin to the underlying fascia. This attachment limits motion and provides a stable platform for grasping and manipulation of objects. In contrast, the dorsal skin is loose with areolar tissue between the skin and extensor tendons. This laxity creates the dorsal subcutaneous space for lymphatic and venous drainage. This structural difference accounts for the dorsal hand swelling seen during infection even when the source is located on the palmar side of the hand **(Bishop et al , 1994)**.

The palmar fascia of the hand provides a stable platform for the skin and protects the underlying structures (Figure 5). This fascia is also the insertion site for the palmaris longus tendon, allowing this muscle to flex the wrist. A septum extending from the palmar aponeurosis to the third metacarpal divides the recesses of the hand into spaces (Figure 6). These spaces are deeper and different from the palmar surface areas described earlier. The thenar space is located to the radial side of the septum and the midpalmar space is situated on the ulnar side. These spaces can be infected primarily or secondarily following flexor tenosynovitis of the digits **(Baratz et al, 2002)**.

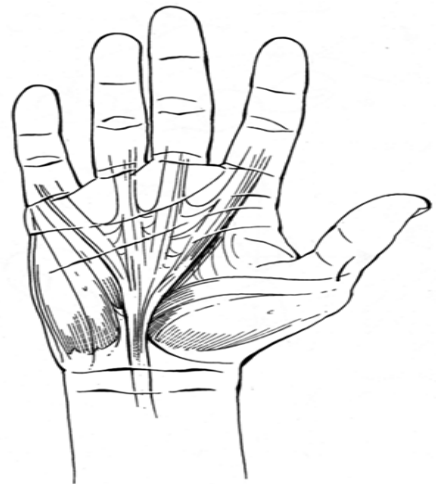

Figure 5: Palmar fascia of the hand provides stability and protection to the hand **(Baratz et al, 2002)**.

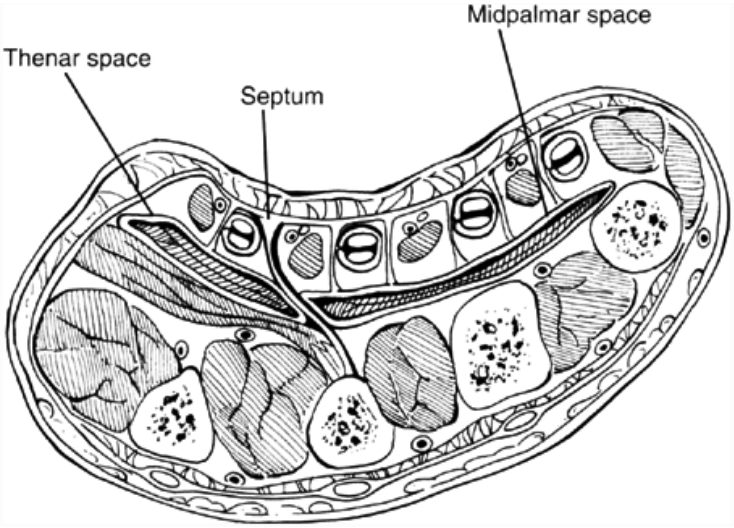

Figure 6: Transverse section demonstrating septum from palmar fascia to third metacarpal, which divides hand into thenar and midpalmar spaces **(Baratz et al, 2002)**.

Blood Supply:

The radial and ulnar arteries supply the hand and digits by a series of arches (Figure 7). The radial artery is located between the brachioradialis and flexor carpi radialis tendons at the wrist. The artery splits into two branches, with the larger dorsal branch coursing under the first dorsal compartment, through the anatomic snuffbox, between the index and thumb metacarpals, and into the recesses of the palm to form the greater part of the deep palmar arch **(Lampe, 1998)**.

A smaller palmar branch travels over the flexor carpi radialis tendon, beneath or through the thenar muscles, and forms the radial component of the superficial palmar arch. The ulnar artery is located lateral to the ulnar nerve at the wrist and adjacent to the flexor carpi ulnaris tendon. The nerve and artery course into Guyon's canal, which is bordered by the pisiform and hook of the hamate. The floor of Guyon's canal is the transverse carpal ligament (TCL) and the roof is the volar carpal ligament (Figure 8). The artery splits into two branches with the larger branch forming the main constituent of the superficial palmar arch. A smaller branch passes deep to connect with the radial artery and form the deep palmar arch **(Baratz et al, 2002)**.