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Variable angle locking technology for mediocarpal partial arthrodesis.
VA-Locking Intercarpal Fusion System 2.4.

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The plates are especially designed for fixation of four corner arthrodesis and feature a low profile design with rounded edges to minimize the risk of soft tissue irritation. They are available in two sizes (Ø 15 mm/6 holes and Ø 17 mm/7 holes) and have K-Wire holes incorporated to allow temporary fixation of the plate.

**Variable angle locking technology**
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**Innovative Instrumentation**
System specific instruments like Reduction Reaming Guides additionally support the surgeon during procedure. The fusion site is fixed during the whole procedure and if desired, compression can be applied with the instrument.
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Dear colleagues

Wishing you a prosperous and happy 2013!
We are proud to bring you another issue of the IFSSH ezine as we start the third year of running this publication for the international hand surgery and therapy community. In this issue you can read about the following:

• Special Feature: The South American Federation of Societies for Surgery of the Hand (SAFSSH). In our endeavour to introduce another member of our international Hand Surgery family, we bring you the History of SAFSSH in this issue. Regional Hand Surgery and therapy groups are important because they not only address regional and local issues, but are also important in keeping us updated as to what our colleagues do in various other parts of the world.
• The IFSSH Committee system has been overhauled and proves to be gaining momentum. It has become prestigious to be involved in producing an IFSSH Committee Report. Presently more than 30 Committees are working on specific reports. As these reports are completed, they are published in the IFSSH ezine and posted on the IFSSH website.

The President and the other Exco members of the IFSSH are conscious of the tremendous amount of work done by the Chairpersons and the Committee Members to prepare these reports. The Hand Surgery Community is indebted to its colleagues who enthusiastically and willingly share their knowledge and experience. We sincerely thank all those who have so far, and will in the future contribute!

You can access all the recent Committee Reports on the IFSSH website: www.ifssh.info
• FINAL ANNOUNCEMENT - Please do not forget to register for the IFSSH Congress in New Delhi in March 2013! The website is www.ifssh-ifsht2013.com

If you miss this Congress, you will miss out on a whole lot!
With sincere regards,

PROF ULRICH MENNEN
President: IFSSH
Editor: IFSSH ezine
1 February 2013

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Obituaries

Professor Renzo Mantero

Renzo Mantero, a pioneer of Hand Surgery and beloved master of many among us, has suddenly deceased on Thursday 1 November 2012.

He was born in Portovenere, Italy, on February 11 1930. He took his degree as Medical Doctor at the State University of Genoa, Italy, in 1954, the Diploma as Specialist in General Surgery in 1959 and the Diploma as Specialist in Orthopaedics and Traumatology in 1962 at the same University. In 1964 he was awarded the title of Professor in Surgery at the State University of Pavia, Italy, and was titular of the Course of ‘Hand Surgery’ at the same university.

He was Chief Surgeon of the Department of General Surgery and Hand Surgery at S. Paolo Hospital in Savona, from 1970 to 1994, and Chief Surgeon of the Regional Center of Hand Surgery in the same hospital from 1995 to 1999, where he was also Emeritus Chief Surgeon and Scientific Director from 2000.

He was a member of the Italian Society for Surgery of the Hand (SICM) from its foundation in 1963, becoming its President in the years 1980-81. He was Member of the Italian Society of Orthopaedics and Traumatology from 1962 and of the major Italian and foreign Societies of Orthopaedics: Foreign member of the GEM (French Society of Surgery of the Hand), Honorary member of the SECMA (Spanish Society of Surgery of the Hand), member of the “Amicale Internationale de Chirurgie de la Main” and many others. He was also a Fellow of the International College of Surgeons (FICS). He was also awarded the status of “Pioneer of Hand Surgery” by the IFSSH at the 2010 Seoul IFSSH Congress.

From 1979 was Director of “Rivista Italiana di Chirurgia e Riabilitazione della Mano e dell’Arto Superiore”, the official journal of Italian Society of Surgery of the Hand. He also was Founder and Director of “Manovre”, a Journal issued by the “Savona Foundation for Hand Studies”, containing studies about the anthropological and cultural aspects of the Hand.

Among his personal studies: Medical Aspects of the Divina Commedia, Hand Symbolism in Christian and Demoniac Culture, The Hands in Verga, Pirandello and Neruda writings; musical researchers on Paganini’s, Robert Schumann’s, Chopin, Liszt and Rachmaninov Hands. Particular studies where performed on Leonardo’s last supper hands, Tiepolo’s hands.

Daniel C. Riordan

Dr Daniel Clifford (Dan) Riordan, 95, passed away Saturday morning, October 27, 2012 in Shreveport, LA, USA. Born October 3, 1917 in Vallejo, CA, USA, he graduated from Stanford Medical School in 1942 and entered the US Army. He was later assigned to Valley Forge Army Hospital in Pennsylvania. In 1945 he became the Chief of Hand Surgery there.

In 1949 Dr Riordan moved to New Orleans, LA, USA and began a lengthy career of hand surgery and teaching, retiring in 1992. He pioneered extensive hand surgery research at the public hospital in Carville. Beginning in 1949 Dr Riordan was board certified and a fellow at the American Academy of Hand Surgery and was elected as president in 1960. The Riordan Hand Society was established in his honor in 1967.

At the Fifth International Hand Surgery Congress in Paris, France in 1992, Daniel Riordan was honoured with the title: “Pioneer of Hand Surgery” by the IFSSH.

With the above credentials, Dr Riordan is the one of the original pioneers in the development of
Hand surgery as it is known today. His love, dedication and enthusiasm for restoring hand function, fuelled his many innovations of surgical procedures and made him an exceptional teacher. “Dan the Hand Man” as he was affectionately nicknamed, influenced many hundreds of prospective Hand Surgeons worldwide.

Acknowledgement is given to Shreveport Times.

Pictured above (year unknown) are the American Society for Surgery of the Hand’s Founder and first President, Sterling Bunnell, MD, (right) and Daniel C. Riordan, MD, who served as the 15th President of the ASSH. Bunnell’s term as President was from 1946-’47, and Riordan’s term was from 1960-’61.
Hand Analogies Survey

Kevin Kruse and John Milton (members of the American Society for Surgery of the Hand) are conducting an interesting survey about the analogies that Hand Surgeons and Hand Therapists use to communicate disorders of the hand and upper extremity to their patients.

Please send us your favourite analogies to ezine@ifssh.info, even if it is only one sentence. We will keep them anonymous, however, if you prefer, we can include your name. Once we have collected your contributions, they will be published in a future IFSSH ezine. This should make very interesting reading!

Below are a few examples:

1. Carpal Tunnel: “There is a ligament that is like a tight rubber band around your nerve.”
2. Hand Infections: “Bacteria like warm, dark, moist places, like the inside of a hand.”
3. Distal Radius Fractures: “The volar plate acts like a rake when holding the bone together.”
4. Flexor Stenosing Tenosynovitis: “Like trying to pull a knotted string through a straw.”
5. Scaphoid Fractures: “This bone is the keystone to the wrist, and when it fails the entire wrist fails.”
6. Dupuytren’s Disease: “A Dupuytren’s cord is like a rope made of scar tissue that shortens and tightens over time.”
7. Stiff Joints: “are like rusty hinges. If the hinges are rusty, changing the doorknob isn’t going to make the door move easier.”
8. Tennis Elbow: “The cortisone does not heal your tendon it merely takes away the pain while your body heals itself.”
9. Post-operative pain medication: “It is like putting out a fire; better to get on it early, than let it get out of control and burn your house down.”
10. Ganglion Cyst: “is like a weak spot on a car tire that gets bigger and bigger.”
11. Flexor Tendon Repair: “Is like gluing two pieces of string together inside a drinking straw. Too much glue and the string can’t slide. Not enough glue and it will pull apart.”
12. Scar Tissue: “is like concrete, easy to work with when it is new but difficult to work with after it has hardened.”
13. Scaph-lunate ligament instability: “This ligament is like the ACL of the knee, but unlike the ACL, in the wrist we haven’t figured out a good way to fix it yet.”
14. Arthritis: “our joints are like tires and you have worn out the tread.”
15. Kienböck’s Disease: “is like someone turned off the water supply to your garden and the crop (the bone) is dying.”
16. SLAC and SNAC wrist: “The scaphoid is the keystone to the wrist and when it fails the entire wrist collapses.”
17. ORIF of hand fractures: “This is like putting a cast under your skin to hold the bone in place while it heals.”
18. Cubital Tunnel: “Your ulnar nerve is getting kinked like a garden hose at your elbow.”
19. Jersey Finger: “When the tendon ruptures it can retract down into your palm like a rubber band.”
20. De Quervain’s: “Is like a rope getting frayed and worn as it is pulled back and forth underneath a rock.”
Dear Colleague,

The second European Hand Injury Prevention Congress was organised and held at the Unfallkrankenhaus in Berlin, Germany on 15 and 16 October 2012. You can find some photos of the congress as well as more information on the official website of Turkish Hand Injury Prevention Society.

We would like to express a special appreciation to R. Böttcher, W. Eichendorf and T. Linz.

Best regards,
Tufan Kaleli
President
Turkish Hand Injury Prevention Society
www.handprevention.org
Dear ‘Hand Family’,

On behalf of the Executive Committee of the International Federation of Societies for Surgery of the Hand I wish you and your families a happy New Year!

Time is flying and another IFSSH Congress is approaching, which means that three more years can be added to our experience and knowledge. Preparations of the 2013 New Delhi Congress commenced many years ago and are overseen by the ExCo of IFSSH. Our friends, the Hand Surgeons of India and especially the Congress Organising Committee, are doing their best to prepare an exciting event, primarily with the scientific programme and, of course, with the enticing social programmes. The number of submitted abstracts is the proof of the international interest in this event and the scientific programme is almost finalised. We are very pleased to share the programmes with our hand therapist colleagues in India and share our experiences and knowledge.

The opportunity to register and book your accommodation directly with the official organisers ensures that you are met at Delhi airport, the transfer to your hotel and the congress venue is organised, and this is all included in your registration fee. Accommodation possibilities are still available in all categories and the Agra Taj Mahal post congress one day tour is a must for all participants.

The Congress will involve several important events. One of these is the Opening Ceremony where IFSSH Pioneers of Hand Surgery from all around the world will be celebrated for their huge contribution to the development of hand surgery. An artistic programme and welcome reception will provide a memorable start for the participants.

The annual Delegates’ Council Meeting will be held on Wednesday 6 March. Our Federation needs the suggestions, input and activity of the national societies and this is forthcoming from their delegates. Our aim is to improve education, teaching and knowledge. New ideas, project suggestions and applications for bursaries and grants are encouraged.

An important act will be the election of the 2019 Congress host and site. Bids have been submitted by the British Society and the German Society. These have been distributed to all national delegates and we encourage your membership to discuss these bids prior to the Delegates’ Council Meeting. If your national delegate cannot attend, it is essential to submit to advise the secretariat (administration@ifssh.info) of an alternate attendee from your society, or submit a proxy vote in writing prior to the meeting. Please also note that only societies who have met their financial obligations are eligible to vote.

With the energetic activity of our President and President Elect, the progress of the Scientific Committees is continuously growing. These reports will be available in Delhi, as well as on the website and published in the IFSSH ezine.

As 2013 begins, it appears that everything is ready for another successful Congress and year of IFSSH activity - our ‘Hand Family’ eagerly awaits your active participation and involvement.

We are looking forward to welcoming you in India!

Zsolt Szabo
Secretary General, IFSSH
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History of the South American Federation of Societies for Surgery of the Hand

By Eduardo R Zancolli III

After World War II, Hand Surgery was performed by only a few individuals in South America. Hand Surgery topics were treated, as in other countries, either by Orthopaedic Societies or Plastic Surgery Societies. With time they tended to develop sub-sections of their national societies for those who had developed an interest in Hand Surgery. There was a considerable reluctance to allow independent Hand Surgery societies to develop.

In 1965 two significant events took place: In Brazil, in July 1965, the first Hand Surgery International Congress was organised under the presidency of Henrique Bulcao de Morales (the Brazilian Hand Surgery Society had already been formed by then). In December 1965 during the SLAOT Congress in Lima, Perù, a group of Hand Surgeons founded the South American Federation for Hand Surgery. The group elected Eduardo A. Zancolli as President (Argentina), Carlos Firpo as Secretary (Argentina), and, Teodoro Ayala (Paraguay) and Enrique Jenkin (Chile) as executive committee members. The other founding members were Edgard Kamel (Venezuela) and Eugenio Elizalde (Perù). The Federation involved the South American Spanish speaking countries which included Argentina, Bolivia, Chile, Colombia, Ecuador, Paraguay, Perù, Uruguay and Venezuela. The second president was Carlos Firpo.

In the 1970’s and 1980’s activity in the South American Federation for Hand Surgery diminished, partly because a variety of national societies were being formed and they tended to concentrate on developing their own national organisation as the highest priority. During this time the president of the South American Federation was Guillermo Loda.

A wide variety of courses and congresses took place in the years that followed and some were held in association with the Brazilian Hand Surgery Society. The Secretary-General of the IFSSH in 1990 was Micky Vargas. The International Federation was somewhat concerned about the viability of the South American Federation and discussions were held in an attempt to develop a regional role for the South American Federation, which at this stage did not include Brazil. Discussions led to a re-launching of the South American Federation (including Brazil) at the South American Congress in Buenos Aires, in 1992 under the presidency of Guillermo Loda. New guidelines were created providing a commitment to organise a Biannual Congress. In the years when there was no congress, an instructional course was to be organised in countries where hand surgery was not as yet well developed. The faculty for such courses (usually between 10 and 20 hand surgeons) would support the course by paying their own transport and hotel accommodation costs.

The tenure of the president was also altered for the rejuvenated federation, and since that time the presidents have been as follows:

1993- Arlindo Pardini (Brazil)
1994-1995- Contreras Gamboa (Venezuela)
1996-1997- Eduardo R. Zancolli III (Argentina)
1998-1999- Walter Albertoni (Brazil)
2000-2001- Alberto Perez (Chile)
2002-2003- José María Rotella (Argentina)
The Federation continues to develop its regional role in South America with its primary concern being the provision of Hand Surgery education to countries in the region where the Hand Surgery services are less developed.

**ABOVE:** Argentine Hand Surgeons during a meeting in 1995 at National Academy of Medicine (invited guest: Ronald Lischeid). 1st row from left to right, Dr Sanguinetti, Dr Lara, Dr Borghi, Dr Archain, Dr Poitevin, Dr Cosentino, Dr Rodriguez Samartino, Dr Ortega, and sitting in front: Dr Nogueira and Dr Ramos Vertiz. 2nd row, from left to right: Dr. Fazzini, Dr Loda, Dr. Dabbah, Dr. Torrano, Dr. Eduardo R Zancolli, Dr Aponte, Dr Lischield, Dr Eduardo A Zancolli, Dr Gamarnik, Dr Miller, Dr Garay, Dr Zaidemberg, Dra Rantica, Dr Cagnone, Dr Rotella, Dr Maquieira.

Recent progress in both clinical genetics and developmental biology has refined our understanding of how limbs develop and malformations occur\(^1\). This brief update highlights our current understanding regarding the genetic basis of congenital anomalies of the upper extremity and hand. This update focuses on upper limb malformations that occur from failed formation or differentiation along known axis-related pathways and utilises the terminology of a recently proposed classification scheme that integrates clinical genetics, developmental biology and recognised upper limb abnormalities\(^2\). According to this classification, failure of axis formation/differentiation may be sub-classified into two groups: failure of the axis along the entire upper limb and failure of the axis within the hand-plate only as shown in Table 1.

The limb can be described in terms of three developmental axes – the proximo-distal axis, the anterior-posterior (or radial-unlar) axis and the dorsal-ventral (or dorsal-volar) axis as depicted in figures 1 and 2. Each of these axes is controlled by signaling centers that initiate a cascade of axis-related pathways.

### Table 1: Selected upper limb malformations related to failure of axis formation/differentiation which will be discussed in the current report

<table>
<thead>
<tr>
<th>Axis Failure</th>
<th>Axis Involved</th>
<th>Malformations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Axis failure along the entire upper limb</td>
<td>Proximo-distal</td>
<td>Transverse deficiency, Amelia,</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Intersegmental deficiency</td>
</tr>
<tr>
<td>Antero-posterior</td>
<td></td>
<td>Radial ray deficiency, ulnar ray</td>
</tr>
<tr>
<td></td>
<td></td>
<td>deficiency, mirror hand</td>
</tr>
<tr>
<td>Dorso-ventral</td>
<td></td>
<td>Nail patella syndrome, WNT7A</td>
</tr>
<tr>
<td></td>
<td></td>
<td>mutation syndromes</td>
</tr>
<tr>
<td>Axis failure within the hand-plate only</td>
<td>Antero-posterior</td>
<td>Radial polydactyly, triphalangeal</td>
</tr>
<tr>
<td></td>
<td></td>
<td>thumb, ulnar polydactyly</td>
</tr>
<tr>
<td></td>
<td>Dorso-ventral</td>
<td>Dorsal dimelia</td>
</tr>
</tbody>
</table>

Failure of the proximo-distal axis (Fig 1): Transverse arrest, amelia, and segmental deficiencies (Table 2):

Limb positioning along the vertebral axis is controlled by Homeobox (HOX) transcription factors upregulating TBX5 in the presumptive upper limb which activates fibroblast growth factor (FGF) 10 in the mesoderm\(^3\). Haploinsufficiency or single allele disruption of TBX5 causes Holt-Oram syndrome with a broad range of upper
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limb abnormalities, although radial defects appear to predominate. In mice, a conditional knockout of TBX5 in the limb stops limb bud initiation and outgrowth. However, the heart is also affected by TBX5 deficiency. Thus, the complete loss of TBX5 function in humans is likely a lethal mutation and no cases have been reported. Mesodermal FGF10 induces Wingless-type MMTV integration site family (WNT) 3 in the apical ectodermal ridge (AER) which upregulates several AER-related FGFs. These AER-related FGFs, particularly FGF8, are the primary signaling molecules of the proximo-distal axis and they maintain FGF10 secretion in the mesoderm. This reciprocal FGF10-FGF8 loop is critical for forelimb outgrowth. Disruption of limb induction or the initiation of AER-related FGFs disrupts the proximo-distal axis and causes transverse deficiencies the most severe form being Amelia, the most severe form of transverse deficiency.

Interrupting the reciprocal FGF loop is also associated with failure or interruption of limb formation. Complete loss of WNT3 function (OMIM 273395) causes tetra-amelia. In animals loss of Fgf10 function or ectodermal FGFs or transcription factors associated with AER formation (such as p63) has also been shown to abate limb outgrowth and lead to a range of defects from tetra-amelia to terminal truncations. More recently, Al-Qattan’s group showed that some patients with complete loss of WNT7A

---

**Table 2: The genetic basis of malformations secondary to failure of the proximo-distal axis**

<table>
<thead>
<tr>
<th>The Malformation</th>
<th>Genetic Basis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Amelia</td>
<td>Disruption of limb induction:</td>
</tr>
<tr>
<td></td>
<td>- Conditional knockout of TBX5 in mice</td>
</tr>
<tr>
<td></td>
<td>- Complete loss of WNT3</td>
</tr>
<tr>
<td></td>
<td>- Complete loss of FGF10</td>
</tr>
<tr>
<td></td>
<td>- Complete loss of p63</td>
</tr>
<tr>
<td></td>
<td>- Some cases of complete loss of function of WNT7A</td>
</tr>
<tr>
<td>Transverse</td>
<td>From animal models, disruption of the Apical ectodermal ridge (AER) or AER-related fibroblast growth factor signaling after initial limb bud formation (level of transverse deficiency is dependent on the time of disruption)</td>
</tr>
<tr>
<td>deficiencies</td>
<td>- Disruption of Shox2 (in animals)</td>
</tr>
<tr>
<td>Intersegmental</td>
<td>- Roberts-SC phocomelia spectrum in humans (ESCO2 gene)</td>
</tr>
<tr>
<td>deficiencies</td>
<td>- Thalidomide-induced tetartogenicity (possibly acts via CRBN and FGF8 expression)</td>
</tr>
</tbody>
</table>

---

**Fig 1: The proximo-distal axis: Mesodermal TBX5 induces FGF10 in the mesoderm. FGF10 induces WNT3 in the AER. WNT3 will then induce FGF8 in the AER which will help maintain FGF10; and the antero-posterior axis: SHH is normally loaded posteriorly and controls the development of the ulna and fingers. SHH induces FGF4 in the posterior aspect of AER. FGF4 will also help maintain SHH activity posteriorly. The thumb and radius develop normally under the influence of GLI3R and SALL4 (in the mesoderm) as well as FGF8 in the overlying anterior ectoderm. Note that Fanconi anemia genes which interact with ubiquitin/ SUMO pathway are highly expressed in the AER.**
function (associated with dorsal-ventral patterning) will also have upper limb amelia suggesting a role for this factor in upper limb induction or maintenance.\textsuperscript{11} Transverse deficiencies are uncommon, and in animal models these malformations can be mimicked by AER removal or functional ablation of FGF signaling\textsuperscript{12,13}

Segmental disruption of the limb has been linked to SHOX and SHOX2 genes. The names of these structurally similar paralogs are derived from the initial recognition that the SHOX (short stature homeobox) gene was associated with the mesomelic short stature of Turner syndrome.\textsuperscript{14} Disruption of Shox2 disrupts upper arm and thigh development in animal models, but has not yet been reported in humans.\textsuperscript{15} Both Roberts (pseudothalidomide) syndrome (MIM 268300) and SC phocomelia syndrome (MIM 26900) are caused by mutations of the ESCO2.\textsuperscript{16} Upper limb malformations vary from radial ray deficiency to phocomelia. The ESCO2 genes code for an acetyltransferase which is involved in the regulation of sister chromatid cohesion during the S phase of cell division. Newborns with thalidomide teratogenicity show phocomelia. Ito et al. recently identified CEREBLON (CRBN) as a thalidomide-binding protein. CRBN forms an E3 ubiquitin ligase complex with damaged DNA-binding protein 1 (DDB1) which is important for limb outgrowth and expression of FGF8.\textsuperscript{17}

Failure of the antero-posterior axis

<table>
<thead>
<tr>
<th>The Malformation</th>
<th>Genetic Basis</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ulnar ray deficiency</td>
<td>Deficiency in the normally located SHH (posteriorly)</td>
</tr>
<tr>
<td>Radial polydactyly - tripalangial thumb-mirror hand spectrum</td>
<td>Ectopic anterior expression of SHH (in humans, commonly due to ZRS mutations)</td>
</tr>
<tr>
<td>Ulnar polydactyly</td>
<td>GLI3 mutations (Greig cephalopolysyndactyly, Pallister Hall syndrome, postaxial polydactyly types A/B)</td>
</tr>
<tr>
<td>Radial ray deficiency</td>
<td>- RECQL4 mutations (Rothmund-Thomson syndrome type II, RAPADILINO syndrome, and Baller-Gerold syndrome) - Fanconi anemia genes (Fanconi anemia and VACTREL with hydrocephalus) - TBX5 (Holt Oram syndrome) - SALL4 (Duane radial ray syndrome) - SALL1 (Townes-Brocks syndrome) - Microdeletion of 1q21.1 (Thrombocytopenia-absent radius syndrome)</td>
</tr>
</tbody>
</table>

Table 3: The genetic basis of malformations secondary to failure of the antero-posterior axis

HOX transcription factors also establish posterior polarity along the radial-ulnar axis which initiates sonic hedgehog (SHH) production from cells that become the zone of polarizing activity (ZPA).\textsuperscript{18} SHH, the primary signaling molecule of the radial-ulnar axis, emanates from the ZPA in the distal ulnar aspect of the developing limb. SHH induces proliferation and ulnarises nearby associated tissues.\textsuperscript{19,20} As shown in Fig 1, the posteriorly located SHH controls the development of the ulna and all fingers except the radial aspect of the index finger. Disruption of SHH or its pathway leads to defects akin to ulnar longitudinal deficiency in animal models.\textsuperscript{21-23}

Normal thumb and radius development only occurs if there is no SHH expression anteriorly. Instead, the development of thumb/radius is under the control of mesodermal TBX5, SALL4, GLI3R, HOXA13, HOXD13 and FGFs in the anterior ectoderm.\textsuperscript{24,25} SALL4 is associated with TBX5 in limb/heart development; and with SALL1 in limb/kidney development. Hence, patients with Duane radial ray syndrome (SALL4 mutation, MIM 607323), Holt-Oram syndrome (TBX5 mutation, MIM 142900), and Townes-Brocks syndrome (SALL1 mutation, MIM 107480) have overlapping features of radial ray deficiency, cardiac and renal defects.\textsuperscript{26} Radial ray deficiency can also be seen with RECQL4 mutations (RECQL4 codes a DNA helicase involved in DNA unwinding) and also with mutations in Fanconi anemia genes\textsuperscript{27} (See Table 3). Al-Qattan\textsuperscript{28}
noted that both genes were involved in ubiquitin-DNA processing and this is interesting because Fanconi anemia genes are highly expressed in the apical ectodermal ridge in which FGF8 directly affects radial ray development. Köppen et al.30 identified a microdeletion of 1q21.1 in a group of 30 patients with TAR (Thrombocytopenia-absent radius syndrome, MIM 274000). In 75% of these patients, the deletion was inherited from an unaffected parent. Köppen et al. concluded that although the deletion is required, it is not sufficient for displaying the TAR phenotype. The phenotype develops only in the presence of an additional as-yet-unknown modifier (named mTAR). Al-Qattan speculated that this mTAR may be related to the ubiquitin pathway. Finally, most cases of VACTERL have been sporadic, but Solomon et al.31 recently reported an increased prevalence of isolated VACTERL clinical features in first-degree relatives.

Ectopic radial expression of SHH has been associated with triphalangeal thumb and preaxial polydactyly.32,33 In animal models, elevated levels of ectopic radial SHH will generate mirror hand or ulnar dimelia.34,35 In humans, most cases of familial radial polydactyly are related to mutations of the ZRS (zone of polarising activity regulatory sequence).32,33,36-39 The ZRS is a long-range limb specific SHH enhancer on chromosome 7q36, approximately 1Mb telomeric of SHH. The ZRS regulates the expression of SHH and point mutations in the ZRS results in enhanced SHH activity and ectopic anterior expression of SHH causing a variable phenotype of preaxial polydactyly and triphalangeal thumb. The racial distribution of the types of thumb polydactyly may also be explained genetically.40 For example; thumb triplications and triphalangeal thumbs are known to be more prevalent in populations with genetic isolates with mutations located at chromosome 7q36. The best example is the South-Western region of the Netherlands where triplication of the thumb and thumb duplication with triphalangism are seen in 8% and 25% of all cases of thumb polydactyly; respectively.41

**Failures of dorsal-ventral axis formation/differentiation axis (Fig 2): Nail-patella syndrome, WNT7A mutation syndromes and dorsal dimelia (Table 4):**

As seen in Figure 2, WNT7A is expressed in the dorsal ectoderm and is the key player of the dorsal-ventral axis, responsible for development of dorsal structures in the hand (such as the nails, the extensor tendons and the thin hairy skin).42 EN-1 expressed in the ventral ectoderm restricts WNT7A to the dorsal ectoderm. WNT7A induces the expression of LMX1B in the dorsal mesoderm and it is also important in maintaining SHH activity in the posterior mesoderm. Loss of EN-1 in animals leads to over-expression of WNT7A ventrally and hence all digits show dorsal dimelia with palmar nails.

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**Fig 2: The dorso-ventral axis: EN-1 is in the ventral ectoderm. It supresses WNT7A preventing its expression ventrally. WNT7A of the dorsal ectoderm induces LMX1B in the dorsal mesoderm and helps to maintain SHH activity.**
as well as dorsalisation of the palmar skin/structures.43 Dorsal dimelia in humans does not involve all digits and probably represents stochastic developmental errors.44 Human dorsal dimelia of the ulnar digits is commonly associated with ulnar-sided malformations such as ‘ulnar cleft hand’.45 Similarly, human dorsal dimelia of the radial digits is usually associated with radial-sided malformations.27,46,47 More recently, Al-Qattan et al.48 identified a different pattern of familial dorsal dimelia which only involved the skin of the proximal palm (all digits were normal in all affected family members). Linkage analysis and exome sequencing in that family identified a heterozygous GLE1 mutation.

Mutations of LMX1B cause nail-patella syndrome with partial loss of dorsalisation evidenced as hypoplastic nails and hypoplastic/absent patellae.49 Finally, WNT7A loss-of-function mutations lead to ventral dimelia (appearance of palmar structures on the dorsum of the hand with absent/hypoplastic nails) as well as ulnar ray deficiency (secondary to loss of the effect of WNT7A on SHH). Humans with WNT7A mutations show a variable phenotype regarding the degree of ventralisation of the dorsum of the hand and also regarding the severity of ulnar ray deficiency. Al-Qattan prefers to call these cases congenital duplication of the palm syndrome50-52 In the genetics literature, the phenotype is known as Fuhrmann syndrome when the defects are mild and Al-Awadi syndrome when the defects are severe.53

Table 4: The genetic basis of malformations secondary to failure of the dorso-ventral axis

<table>
<thead>
<tr>
<th>The Malformation</th>
<th>Genetic Basis</th>
</tr>
</thead>
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<tr>
<td>Dorsal dimelia</td>
<td>- EN-1 mutations in animals</td>
</tr>
<tr>
<td></td>
<td>- Dorsal dimelia in humans are associated with other radial/ulnar malformations</td>
</tr>
<tr>
<td></td>
<td>- One family with dorsal dimelia confined to the proximal part of palm was linked to GLE1</td>
</tr>
<tr>
<td>Nail-patella syndrome</td>
<td>LMX1B haplo-insufficiency</td>
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<td>WNT7A mutations</td>
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Reference List

7 Mariani FV, Ahn CP, Martin GR. Genetic evidence that FGFs have an instructive role in limb proximal-distal patterning. Nature 2008;453:401-405.


Clinical case

Reply to clinical case in IFSSHezine #8 November 2012

CLICK HERE to view case

We have seen similar cases, such as this example below:
A boy, 4 years. According to parents, at a birth the deformation was minimal, but over the years it progressed. The movements at the elbow joint is only slightly limited. The cosmetic appearance is a deformation and shortening of the forearm.

Accompanying pathology: HIV +

Is it a variant of ulnar dysplasia or an HIV-combined bone pathology?

Sincerely yours,
Prof. I. Schvedovchenko.
The Chief of First Children Department of St. Petersburg Albrecht Centre of Disabled Persons
St. Petersburg, Russia
For Linda, a 17 year old girl, who was severely burnt in a motor vehicle accident, painting her own finger nails was a major achievement! Linda sustained extensive burn wounds to her face and head, chest, legs, right arm and hand. She was initially treated in a burns unit and finally discharged from hospital four months after her injury. This brief case presentation will focus on the rehabilitation of the right dominant hand and highlight her achievements over a period of six months.

I first met Linda five months post injury. Her hand was covered with dressings and she did not allow me to touch it. Linda was finally persuaded to allow me to remove the dressings so that I could see for myself what the hand looked like. A number of open wounds were evident, the web spaces were narrowed, the entire hand was extremely sensitive and very painful to touch. All the fingers were in a flexed position, had been amputated at the level of the DIP joints and a full thickness skin graft covered most of the dorsum of the hand. I was not allowed to touch the hand at all, so we arranged for a full wound inspection and re-application of dressing under general anaesthetic in theatre.

After the new dressings were applied, a splint was made to try and maintain the maximum extension of the fingers and the first web space.

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Painting her own finger nails...
Reduced wound dressings
Besides the general wound care, soft tissue management and splinting, we concentrated on functional use of the hand and soon persuaded Linda to use her hand in functional activities.

Examples are seen in the photographs below.

Starting leatherwork

Doing leatherwork using both hands

Close-up showing lateral pinch
The wounds on the fingers healed and she only had an open wound on her forearm. Splints were used to try to obtain better abduction/opposition of the thumb, in an attempt to improve her hand function. In order to achieve a better force application, a dynamic splint was tried out. Unfortunately this splint did not work very well, since it caused too much discomfort and we returned to static splinting.

Using a light-weight clamp to pick up pieces of a game

Picking up a ball

A dynamic thumb abduction splint

Back to static splinting
Linda was slowly using her hand more, but was severely hampered by the decreased extension of the fingers and the narrow web spaces. The dorsal full thickness flap, which was used to cover the large skin defect of the dorsum, covered the proximal aspects of her fingers as well, which resulted in an inability to abduct the fingers. It was decided to implement the first phase of the reconstructive surgery. The aim of surgery was to increase extension of the fingers and increase the web spaces. It was also important to re-position the thumb so that she would be able to achieve some opposition and possibly reach the tips of the other fingers.

The hand from the front

Release of the flexion contractures

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Learning points

Here are the most important points I learnt from treating Linda:

- The so-called ‘safe position’ of the MP joints (90 degrees flexion) is not functional. In Linda’s case the flexed position of the MP joints stopped her from using the hand, as she could not open the fingers wide enough to grasp an object. It was only when the contractures were released, that she could start using the hand to pick up and hold something. It is therefore important to maintain the MP joints in flexion for no longer than two to three weeks after a burn or skin graft.

- A positive and encouraging approach is the most valuable asset a therapist and family members can have. This helps a burn victim much more than doing things for him/her.

- Simple, almost child-like activities, such as building a tower with lightweight blocks can be quite challenging for anybody, regardless of age. Provided the activity is presented in a challenging way. In Linda’s case, the first activity she did after the reconstructive surgery was to build a tower with lightweight sponge blocks of varying shapes. Once she could accomplish this activity the rest followed. Exercises and activities should always be pitched at a slightly higher level than the patient thinks he/she can do.

- Participation in meaningful activities still remains one of the best ways of obtaining patient cooperation and independent action. Once she could do one or two activities, she lost the fear of movement and allowed me to touch and gently move her hand. This also paved the way for her to start thinking about other activities she could do. She then tried some of these out at home.

- Although Linda was only referred to me five months after the accident and her hand was in a fixed position, with very little movement, a combination of surgery and therapy made the difference to her ability to do some activities with her affected hand. This would not have been possible if the surgeon, therapist, wound care specialist and the family members were not able to work together as a team.

- Lastly, the resilience of patients who have suffered from severe injuries and survive, never ceases to amaze me. With a lot of gentle support from those around them, they can slowly start making a life for themselves again, a life that is sometimes changed beyond recognition.
Opening the bottle of nail varnish by herself

Good control

Painting her sister’s nails
Every small achievement in Linda’s rehabilitation process was a major step in the right direction. She began to experiment with other activities and enjoyed creative activities and activities that posed a particular challenge for her.

The new web-space is large enough to hold a small glass bottle

Building with blocks

Writing with the ‘new hand’

Palm six weeks post-surgery

Trying a new activity
Holding left and cutting right

Trying a new paint technique
It is nearly a year since the accident and Linda is still gaining function every week. Her greatest wish is to be able to tie up her own hair. This still requires much practice, but with perseverance, she should be able to do this activity soon.

Although this short article does not represent all the treatment Linda received over the past six months, it highlights aspects of her rehabilitation programme that were particularly challenging for her. She passed each ‘test’ with flying colours.

We look forward to the day when she is fully rehabilitated and she is able to use the injured right hand to its full capacity. She has learnt many new skills with her left hand and it will remain that way. Her left hand will be the ‘dominant’ hand and the right will be the supporting hand for all activities that require bilateral hand function. Linda has recently started driving and is coping very well. She certainly has come a long way since the first time she walked into my practice, with the hand covered with bandages, forearm in supination, wrist in neutral and marked ulnar deviation. She did not allow anybody to touch the hand and struggled to think that treatment would not be painful, as she had been through so much during the preceding five months.

I am proud of her achievements and I salute her!
Prevention of flexor pollicis longus tendon rupture after volar plate fixation of distal radius fractures

Japan

“regardless of which plate should be used and where the plate should be fixed, all cases are susceptible to correction loss, which has been reported as a factor of tendon rupture”

A recent study published in Hand Surgery evaluated the presence of tendon irritation of flexor pollicis longus (FPL) for cases of distal radius fracture treated with volar plates to prevent FPL tendon rupture.

Dr Kaoru Tada, from the Department of Orthopaedic Surgery, Kanazawa University Medical School, explained the main aim of the article is to prevent FPL tendon rupture for cases of distal radius fracture treated with volar plate.

“The results of our study indicate that ‘FPL tendon irritation’ is likely to appear before tendon rupture. Therefore, FPL tendon rupture might be prevented by plate removal in patients who complained of tendon irritation,” Dr Tada added.

According to Tada, the most important outcomes of the research are that regardless of which plate is used and where the plate should be fixed, all cases are susceptible to correction loss, which has been reported as a factor of tendon rupture. “Consequently, we believe that evaluation of FPL tendon irritation during the follow-up period is a rational way to prevent FPL tendon rupture,” he said.

Tada and his team are undertaking further investigations to check FPL tendon irritation for cases of distal radius fracture treated with new plates that are designed to set more proximal than earlier plates. Moreover, it is under evaluation about the sensitivity and the specificity of the test about FPL tendon irritation.

JOURNAL REFERENCE
Hand Surgery, Vol. 16, No. 3 (2011) 271 275
The importance of shoulder external rotation in activities of daily living: improving outcomes in traumatic brachial plexus palsy

Dr Angela Wang, Associate Professor of Orthopaedic Surgery at the University of Utah, recently published a study about the importance of shoulder external rotation in activities of daily living in normal individuals.

“The main goal of our article was to more closely examine the role of shoulder external rotation in normal subjects in activities of daily living, and extrapolating that data to see if possibly focusing on restoring external rotation in adult brachial plexus injuries would be beneficial,” Dr Wang explained.

In the study, 31 normal individuals performed 12 common activities of daily living (ADLs) wearing a custom shoulder orthosis designed to selectively limit shoulder external rotation to three different settings.

Wang noted there were several significant outcomes. “We were interested to learn that activities closer to the face were much more difficult with more limited external rotation. We think that shoulder forward elevation plays a bigger part in these activities. Activities at or below the waist had surprisingly little limitation despite loss of external rotation. Also, even restoring a little external rotation had a big effect in improving ability to do tasks,” Wang added.

The results support the notion that restoring shoulder external rotation in the treatment of traumatic brachial plexus palsy patients might improve outcomes by decreasing patient disability and increasing the ability to perform ADLs.

When asked about her future research in this area, she said: “We hope to emphasise that shoulder external rotation plays a large part in ADLs, possibly as much as forward elevation, which has been more traditionally emphasised as a goal in restoration of shoulder function. We hope to be able to compare the two shoulder functions in the future and perhaps determine the relative importance of one over the other.”

JOURNAL REFERENCE
Journal of Hand Surgery
Volume 37, Issue 7, Pages 1430-1436, July 2012
Marc Iselin was born in 1898 and died 20 November 1987.

In 1928 Iselin wrote a monograph on trauma and infections of the hand, with a new edition in two volumes in 1939, which was for many years the basic French reference. He also made an important contribution to the modern surgery of the lung.

Since 1955, his main activity was devoted to Hand Surgery and he developed in his Surgical Unit in Nanterre, France, a dedicated centre for the practice and teaching of Hand Surgery. Twice a year, one-week courses were organised and attended by surgeons from all over Europe and South America, until he retired in 1964.

He supported the GEM since its creation in 1964 and has been one of the early presidents and one of the most active and stimulating members. After he retired from his hospital, he continued his international teaching, organising courses in Italy and South America.

Marc Iselin was a man with a great personality and firm knowledge. His surgical indications when confronted by complex problems showed a philosophical approach towards life, above pure cold surgical techniques. His presence was imposing. In his book "L'adventure en Chirurgie" (1978) he reflects on 'life', which is forever changing, as well his successes, his deeper inner feelings and his struggles.

The courses at Nanterre brought together doctors from different parts of the world, year after year. They dissected anatomical preparations and absorbed the teachings of this master, which constituted true learning. In addition to this surgical teaching-learning experience, he also taught the principles of rehabilitation and splint making. His name will remain closely attached to the French and international history of Hand Surgery.

Kauko Juhani Vainio was born on May 1, 1913, in Sääminki, Eastern Finland. Upon graduating from medical school in 1939, he was immediately drafted for war service as a physician. Following training at the Departments of Surgery and Orthopedics at the University Central Hospital of Oulu, Helsinki, and Turku, he was certified as a specialist in surgery and orthopedics in 1951. Upon his return from a year of study in the United States in 1952, he was appointed Director of Orthopedics at the Rheumatism Foundation Hospital, where he remained until his retirement in 1975. He was appointed associate professor in 1956 and professor in 1970.

The most prominent part of Vainio’s life work was undoubtedly at the Rheumatism Foundation Hospital, where he closely cooperated with Professor Veikko Laine, the internist. The two specialists developed a holistic team approach to the treatment of rheumatoid arthritis. At the time, however, the orthopedic treatment of this condition was an entirely new concept, and the two pioneers met with fierce resistance. Strong willed, the two-man team eventually turned a humble beginning into outstanding achievements that benefitted many severely handicapped patients.

Orthopedic treatment yielded positive results from the very beginning, and as early as the 1950s, the team’s findings published at international meetings and in scientific journals received widespread attention and respect. Assiduous research and the development of new therapeutic methods put Heinola on the world map as the Mecca of treatment, notably surgery, for rheumatic diseases. Kauko Vainio became known as the father of systematic rheuma-orthopedics, and he attracted almost a thousand specialists in rheumatology to Heinola from all parts of the world. His reputation as an inspiring teacher quickly spread and won him popularity on all continents. Gifted in languages, he always tried, as far as possible, to lecture and communicate in the local tongue.

Kauko Vainio participated enthusiastically in specialist organisations in both Finland and abroad. He was President of the Finnish Orthopedic Society from 1961 to 1973, and was awarded honorary memberships in 11 surgical, rheuma-orthopedic, and hand-surgery societies, and in the rheumatological societies of seven countries. He received the Finnish Matti Ayräpää medical award in 1970 for his achievements.

Kauko Vainio’s proficiency was not limited to medicine only; he was a keen student of birds, plants, and particularly mushrooms; and he endeavored to grow various mushrooms in his garden. When travelling, he always took time to find out about the local environment, and he enlivened his travel reports with beautiful photographs and expert descriptions. Even after his retirement, he continued to cherish long-term patient contacts. He was particularly active in keeping abreast with the literature, remaining in touch right up until his death in 1989.

Martti Hämäläinen
Director of Orthopedics
The Rheumatism Foundation Hospital, Heinola, Finland

The IFSSH ezine acknowledges with thanks the above information obtained from Acta Arthop Scand 1989; 60(4)505-506
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- Decision Making for Partial Carpal Fusions – Gregory Ian Bain, Duncan Thomas McGuire
- Scaphocapitolunate Arthrodesis and Radial Styloidectomy: A Treatment Option for Posttraumatic Degenerative Wrist Disease - Melissa Klausmeyer, Diego Fernandez
- Long-Term Results after Midcarpal Arthrodesis - Florian Neubrech, Marion Mühldorfer-Fodor, Thomas Pilla, Jörg van Schoonhoven, K. J. Prommersberger
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- Long-Term Outcomes of Proximal Row Carpectomy: A Systematic Review of the Literature - Harvey Chirm, Steven L. Moran
- Wrist Arthroscopy under Portal Site Local Anesthesia (PSLA) without Tourniquet - Michael T. Y. Ong, P. C. Ho, Clara W. Y. Wong, Sally H. S. Cheng, Wing-Lim Tse
- A Novel Technique for Detecting Instability of the Distal Radioulnar Joint in Complete Triangular Fibrocartilage Complex Lesions - Florian Hess, Mazda Farshad, Reto Sutter, Ladislav Nagy, Andreas Schweizer
- Total Wrist Replacement: A Retrospective Comparative Study - William Cooney, Jennifer Manuel, John Froelich, Marco Rizzo
- Combined Perilunate and Axial Ulnar Dislocation of the Wrist - Robert Gvozdenovic, Niels Søe Nielsen, Marc Garcia-Elias
- Case Report of Long-Term Results of Bixial and Volz Total Wrist Arthroplasty - Scott R. Ekroth, Frederick W. Werner, Andrew K. Palmer
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A Carpal Ligament Substitute Part 1: Polyester Suture – John A. Martin Jr, MD, Marwan A. Wehbé, MD

A Carpal Ligament Substitute Part 2: Polyester Suture for Scapho-Lunate and Triquetrolunate Ligament Restoration – Marwan A. Wehbé, MD, Mary Lee Whitaker, MD

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12th Triennial Congress of the IFSSH &
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March 17-18, 2013
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www.miamihandcourse.com

Day 1: Sunday, March 17th, 2013
Didactic Course (includes lunch and evening dinner event)

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For more information including course and lab agenda, faculty, accommodations info, and more, please see MiamiHandCourse.com

Advanced Derby Hand Course
23-24 May 2012
Derby, United Kingdom
www.pulvertaftandcentre.org.uk

The Pulvertaft Hand Centre, Derby, UK is hosting this comprehensive two-day conference for Hand Surgeons, including: Case based discussions, tips and hints on technique from the experts, current concepts in hand surgery, controversial management issues. Leading national and international Faculty will debate topics including: Dupuytrens Contracture, the Wrist, trauma, flexor tendons, peripheral nerves, joint replacement, CMCJ thumb.

FESSH Congress 2013
29 May – 1 June 2013
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www.fessh2013.com

The 18th FESSH Congress will be held at the capital city of the Turkish Riviera, Antalya between 29 May and 1 June 2013. Antalya is located on the southwestern coast of Turkey. The region is famous for its historical heritage, blue sea, warm weather and is accessible with direct flight from European cities. Detailed information about the congress is available on www.fessh2013.com.

Arthroscopy and arthroplasty of the wrist
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www.sicm.it/norme_editorialien.html

The course is designed for specialists in hand surgery, orthopaedics and plastic surgery who want to improve their technical skills in the diagnosis and treatment of wrist disease. Experts will take lectures and presentations followed by arthroscopic and open surgical techniques on anatomical specimens. Sessions will consider clinical diagnostics for each form of instability or other pathologies of the wrist. Open and arthroscopic techniques will be presented in details with their specific indications. Each participant will bring clinical cases to discuss with experts and will have at least one anatomical specimen.