

ORIGINAL ARTICLE

An Overview of Treatments for Congenital Forearm Longitudinal Deficiencies

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Summary

Introduction. Congenital anomalies are rare, affecting 1-2% of new-borns and only 10% of them have upper limb defects. Incidence of forearm longitudinal deficiencies is 1:55 000-1:100 000 live births which means that 1 case in every two to four years can be expected in Latvia.

Centralisation surgical procedure was the main treatment option until 2008, but it had the worst functional outcome (as literature data also show).

Starting from 2010, several new surgical techniques like microvascular metatarsophalangeal joint transfer, epiphyseal fibula flap transfer and wrist radialisation had been performed in the Microsurgery Centre of Latvia.

Aim of the study. A retrospective study to collect and assess functional outcomes of different types of surgeries.

Material and Methods. 5 patients (7 hands) with radial club hand deformity, one ulnar club hand deformity and one ulnar dimelia were treated in the Microsurgery Centre of Latvia from 2012 to 2014. The Paediatric Evaluation of Disability Inventory (PEDI) was used to evaluate functional outcome in children. ROM (range of motion), pinch and grasp forces were measured to establish wrist motion and stability. Visual analogue scale (VAS) was used to evaluate parents' satisfaction.

Results. 3 patients (5 hands) were selected for the trial. Two patients (3 hands) were not evaluated as thumb reconstruction is not done yet. Patients had a small (10-25°) reoccurrence of radial deviation after centralisation and radialisation procedures, whereas MTP (metatarsophalangeal) joint transfer patients had straight wrists. Wrist joint functionality in MTP patients was following: extension of 10°-35° and flexion of 20°- 90°. In MTP transfer patients, PEDI score was 56-58 for daily activities (normal score – 50), 69-70 for mobility (normal score – 63). VAS score was greater for MTP joint transfer patients.

Conclusion. Radial and ulnar deficiencies are rare and reconstructions should be done at age 2 - 4 years, because late reconstructions are more complicated due to the stiffness of the radial fingers. Radialisation procedure is the first choice for wrist stabilization, but, in some cases, MTP joint transfer or vascularised fibular head transfer can provide a better functional outcome in radial club hand patients.

Key words: radial club hand, vascular epiphyseal fibula, metatarsophalangeal joint transfer, ulnar dimelia

INTRODUCTION

The classification of congenital hand and upper limb anomalies was proposed by Frantz and O'Rahilly and presented by Swanson in 1968 (5). It was adopted in a revised form by the International Federation of Societies for Surgery of the Hand (IFSSH) in 1984 (Table 1). New classification was proposed by Oberg, Manske and Tonkin in 2010 based on current understandings of limb development along the 3 axis (Table 2).

Most congenital upper limb anomalies occur in the embryonic period from 3rd to 7th week (5). Radial club hand or radial longitudinal deficiency can be found in 1:30 000 – 1:100 000 new-borns. Radial longitudinal deficiencies monolateral or bilateral club hands occur as isolated deformities without any other malformation or as a part of a syndrome, the most frequent of which are Fanconi anaemia; Holt-Oram syndrome (in combination with congenital heart defects); TAR syndrome or thrombocytopenia - absent radius, in which the thumb is always presented; and the VATER association or VACTERALS association (vertebral anomalies, anal atresia, cardiac defects, tracheoesophageal fistula,

oesophageal atresia, renal defects, limb deformities, single umbilical artery) (3). Hypoplasia or absence of the radius has been divided into four grades by Bayne. James, McCarroll and Manske have added an additional category (10) in which the radius is fully formed but with a radial deviation of the wrist (Table 3).

Ulnar deficiency of the forearm or ulnar club hand is much less common than the occurrence of radial club hand. Classification for ulnar club hand deformities was described by Bayne and was based on radiological findings later updated by Cole and Manske (Table 4). Surgical treatment is required for type II-IV to stabilize wrist and elbow joints (14).

Mirror hand (ulnar dimelia) is an extremely rare congenital anomaly of the upper limb. There are less than 100 cases described in literature. Mirror hand is a true duplication of the ulna; the radius and radial fingers (thumb) are not present (2;6). Usually poor flexion of a stiff elbow joint (25-30 degrees) is present (16). Thumb has to be created from extra ulnar digits.

AIM OF STUDY

The aim of presented research was to collect and assess functional outcomes for radial-ulnar (anterior-posterior) axis formation/differentiation deformities using different types of surgeries.

MATERIAL AND METHODS

5 patients (7 hands) with radial club hand deformity, one ulnar club hand deformity and one ulnar dimelia were treated in the Microsurgery Centre of Latvia from 2012-2014 (Table 5).

All patients had Litos® external fixator for soft tissue distraction before the wrist stabilisation. Two patients (3 hands) with grade IV (Bayne) underwent 2nd MTP joint transfer at the age of 2 - 4. A male patient with TAR syndrome and involvement of both hands was operated using 2nd MTP joint transfer: right hand at the age of 32 months, operation time – 4h; left hand at the age of 41 months, operation time – 3h 5 min. A female patient with right hand deformity of grade IV (Bayne) and with thumb hypoplasia of grade IIIc (Blauth) (15) was operated at the age of 37 months using 2nd MTP joint transfer. Thumb reconstruction with a 2nd toe to hand transplant was done at the age of 54 months. A new modified technique was used for thumb stabilization – metatarsophalangeal joint arthrodesis, which provides a stability and straightness of the metacarpal bone (Figure 1).

One centralisation was performed to stabilise a wrist joint for a 12 years old girl, as she did not tolerate external fixation. A boy (two hands) had a radialisation procedure at the age of 3 and 4 with following pollicisation of the three digital hand. The patient had Holt-Oram syndrome and loss of the left humerus head. The humerus head was reconstructed with a vascularised fibula proximal head transplant at the age of 2.

A vascularised fibulae head transfer was performed for a patient with grade III (Bayne) deformity at the age of 10 (Figure 2).

A patient with grade IID ulnar club hand deformity was treated in the Microsurgery Centre of Latvia in 2013. C arm deformity was corrected at the age of 6 months using open wedge osteotomies for dividing radius and ulnar constriction bands. A thumb was created using one of three digits; extra digit without metacarpal base was used as pedicle flap and transposed to the ulnar side of the wrist joint at the age of 12 months (Figure 3).

A patient with ulnar dimelia or mirror hand was treated in the Microsurgery Centre of Latvia in 2014. Patient had duplicated ulna and 7 digit hand with no thumb and no active motion in the elbow joint. "Radial head" of the ulna was removed at the age of 6 months with following active splinting during next 3 months and 45 degree passive flexion was achieved. Hand operation to create a thumb was done at the age of 14 months. A new designed surgery – "finger switch procedure" – was performed. A hand consists of 7 digits – 4 at the ulnar side and 3 at the radial side, all three radial digits had 3 phalanges with some movements preserved. The second radial finger had a duplication of the distal phalange

and delta phalange; therefore, during the surgery, the second ulnar digit was transposed to the thumb with arthrodesis in the DIP joint and the first radial digit was transposed to the second finger position. All digits were transposed as pedicle flaps. Two extra radial digits were removed (Figure 4). The Paediatric Evaluation of Disability Inventory (PEDI) was used to evaluate the functional outcome in the children. ROM (range of motion), pinch and grasp forces were measured to established wrist motion and stability. VAS was used to evaluate parents' satisfaction.

Anaesthesia methods: After premedication with Midazolam 0.5 mg/kg intranasally using Mucosa Atomization device, half dose of in each nostril, general anesthesia was done. Induction was achieved with 50% Nitrous oxide, Oxygen and sevoflurane with a face mask, and LMA or endotracheal tube was inserted for airway management. During maintenance Fentanyl 2-3 mcg/kg and sevoflurane was used until 0,9 MAC. Peripheral plexus brachialis or wrist block using 0.25% of Naropin® solution 1.25 mg/kg was performed in all cases for postoperative pain relief. Monitoring consisted BP, HR, ECG, SpO₂, BIS, ETCO₂.

RESULTS

3 patients (5 hands) were included in the trial. Two patients (3 hands) were not evaluated as thumb reconstruction is not done yet. Patients after centralisation and radialisation procedures showed small (10-25°) reoccurrence of radial deviation. But all MTP patients had straight wrists. Wrist joint functionality in the MTP patients was following: extension of 10°-35° and flexion of 20°- 90°. In the MTP transfer patients, PEDI score was 56-58 for daily activities (normal score – 50), 69-70 for mobility (normal score – 63). VAS and PEDI was better in the patients after MTP transfer (Table 6).

DISCUSSION

Pathologies of radial-ulnar (anterior-posterior) axis formation/differentiation are rare. Several reconstruction methods have been described over time. According to the OMT classification, pathologies of radial-ulnar (anterior-posterior) axis formation/differentiation are radial longitudinal deficiency, ulnar longitudinal deficiency, ulnar dimelia, radioulnar synostosis, congenital dislocation of the radial head and humeroradial synostosis (15).

Surgical treatment of radially deviating hand with a bone graft and forming Y-form distal ulna was published by Albee in 1928, who used a simple tibia bone (1). Starr reported a non-vascularised fibula epiphysis transfer after 6 weeks of preoperative skeletal tractions in 1945 (7). They had multiple causes of failure, including disruption of the ulnar growth plate and subsequent increase in limb-length discrepancy, inadvertent ankylosis or arthrodesis of the wrist, loss of motion and failure of the transplanted bone to grow with eventual loss of radial support (18).

There are two most common reconstructive options - centralisation and radialisation (10). During microsurgical era, second metatarsophalangeal joint transfer was suggested for wrist stabilization. Only few articles can be found using fibula proximal epiphysis microvascular transfer for trauma patients (17).

Centralisation of the wrist on the ulna is the standard treatment to correct a radial deviation. The principle of the centralisation procedure is simple - the head of the ulna is placed under and in the central carpal bones (9). This procedure is performed in patients at the age of 1. Surgery at this time allows to improve the length of the forearm and provides a foundation for the development of motor function within the hand. Unsatisfactory results due to damaging ulnar growth plate in his series of centralisation between 1969 and 1979 were published by Buck-Gramcko (3).

Radialisation procedure was described by Buck-Gramcko and published in 1985. The main difference from centralisation is that the head of the ulna is positioned under but not in the centre of the radial carpal bones. This leads to better mechanical advantages, because the lever of the arm is much longer to the ulnar side and more powerful by the radial positioning of the ulnar head and transposition of the radial wrist muscles. The mobility is better, because no carpal bones have been excised. Further result improvement can be obtained through preoperative distraction of soft tissue (2). Long term follow-up studies showed better forearm stability and growth compared to the results of a centralisation procedure (3;13).

The surgical treatment principles for the stabilization of the hand into a correct position with a vascularised second metatarsophalangeal (2nd MTP) joint graft was developed by Simmo Vilkki in 1987 (19). Long term follow-up results published in 2008 showed that the average length of the ulna was 15,4cm (20-10cm) and the total active wrist motion was 83 degrees (30-115°) which were better than the results reported in the long-term studies of centralisation and radicalisation (8). Foot problems are always a question in case of a vascularised

toe or joint transplantation, but long term studies showed that there are excellent results to be expected (11). Thumb reconstruction has to be performed for patients with thumb hypoplasia soon after the wrist stabilization to achieve pinch and grasp functions.

From 2010 until 2014, all main reconstructive methods have been used in the Microsurgery Centre of Latvia to reconstruct radial club hand deformities. A radialisation procedure is the first choice for wrist stabilization, but in some cases MTP joint transfer or vascularised fibula head transfer can provide a better functional outcome for radial club hand patients.

Ulnar club hand deformity is similar to radial club hand, but, usually, the wrist joint is stable. C arm deformities should be treated at an early age using splints and osteotomies.

Ulnar dimelia or mirror hand deformity is very rare and variable therefore there are no standardized procedures. Finger switch procedure is hard to manage, but good aesthetic and functional outcomes can be achieved. The most difficult part is stiffness of the elbow joint. It looks like additional muscle transposition or functional gracilis muscle transfer is required to achieve active movements. Thumb reconstruction is great challenge in patients of longitudinal deficiencies and pollicisation is first procedure to manage but toe to hand transfer can be used for some patients to achieve good aesthetic and functional outcome.

CONCLUSION

First results show that wrists are more stable and there are no reoccurrences of radial deviation, if a vascularised second metatarsophalangeal joint is used for reconstruction. For some patients with Bayne grade II and III deformities vascularised fibula head transplant can be used to stabilize the wrist. Reconstructions should be done at the age of 2 – 4 years, because late reconstructions are more complicated due to the stiffness of radial fingers.

Conflict of interest: None



Fig. 1. 1A. Patient with radial club hand deformity IV (Bayne). 1B distraction with external fixator. 1C 1 year follow up after 2nd MTP joint and 2nd toe to hand transfer.

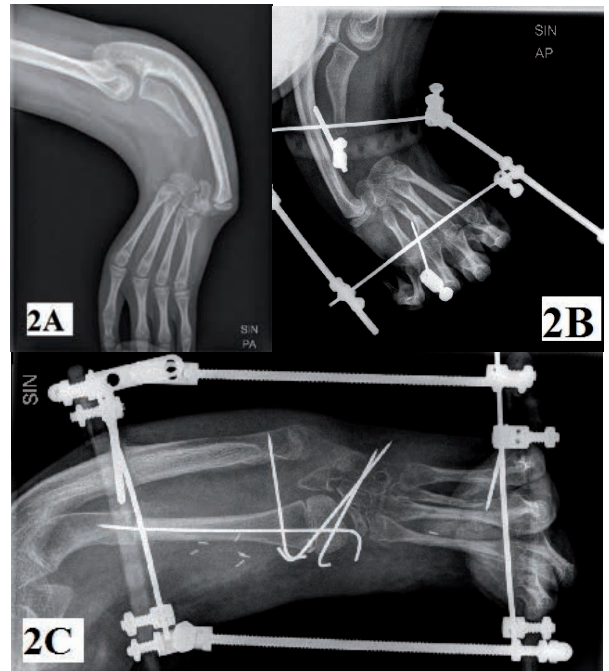


Fig. 2. 2A 10 years old girl not treated radial club hand deformity grade III (Bayne). 2B Litos® external fixator applied. 2C 2 month after fibula head transfer.



Fig. 3. Patient 3 month of age. 3A Ulnar club hand type IID with 'C arm deformity'. 3B right hand after correction of radial deformity no ulnar deviation preserved. 3C Thumb reconstruction and wrist stabilization using 'spare part' surgery. 3D Reconstructed hand 3 month after 'spare part' surgery.

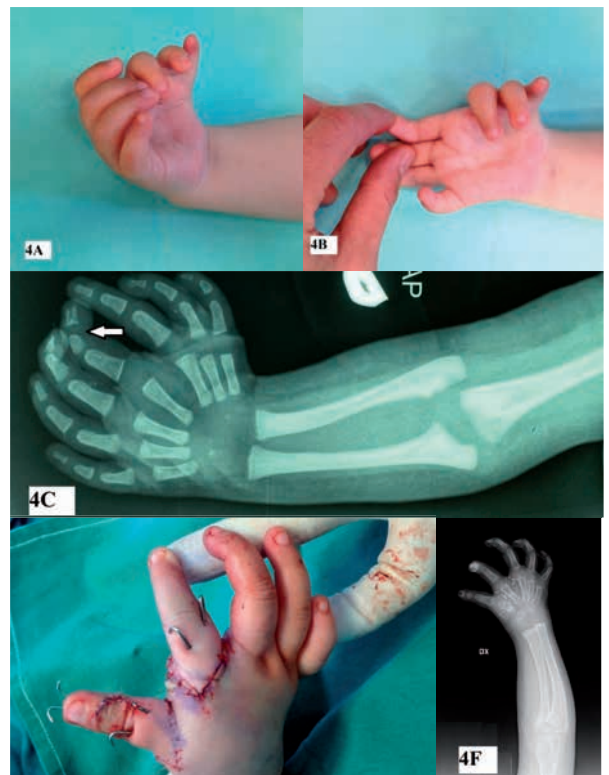


Fig. 4. Child with ulnar dimelia or mirror hand deformity. 4A – 4B seven finger hand with movements presented for radial digits. 4C X ray picture with two 'ulnas' and no radius, triphalangeal digits with duplication and delta phalange of second ulnar digit. 4D Result after 'Finger Switch Procedure'. 4E X ray 6 month post op.

Table 1. Embryologic Classification of congenital Malformations (Swanson 1968).

I. Failure of formation of parts
A. Transverse deficiencies
B. Longitudinal deficiencies
II. Failure of differentiation
A. Synostosis
B. Radial head dislocation
C. Symphalangism
D. Syndactyly
E. Contracture
III. Duplication
A. Thumb
B. Triphalangism/hyperphalangism
C. Polydactyly
D. Mirror hand
IV. Overgrowth
A. Limb
B. Macrodactyly
V. Undergrowth
VI. Congenital constriction band syndrome
VII. Generalized skeletal abnormalities

Table 2. OMT Classification of Congenital Hand and Upper Limb Anomalies (Oberg, Manske, Tonkin 2010).

I. MALFORMATIONS	
A. Abnormal axis formation/differentiation—entire upper limb	
1. Proximal-distal axis	
2. Radial-ulnar (anterior-posterior) axis	
i. Radial longitudinal deficiency - Thumb hypoplasia (with proximal limb involvement)	
ii. Ulnar longitudinal deficiency	
iii. Ulnar dimelia	
iv. Radioulnar synostosis	
3. Dorsal-ventral axis	
4. Unspecified axis	
B. Abnormal axis formation/differentiation—hand plate	
1. Proximal-distal axis	
2. Radial-ulnar (anterior-posterior) axis	
3. Dorsal-ventral axis	
4. Unspecified axis	
I. DEFORMATIONS	
A. Constriction ring sequence	
B. Trigger digits	
C. Not otherwise specified	
II. DYSPLASIAS	
A. Hypertrophy	
B. Tumorous conditions	
III. SYNDROMES*	
A. Specified	
B. Others	
*The specified syndromes are those considered most relevant; however, many other syndromes have a limb component categorized under “B. Others”.	

Table 3. Global Classification of Radial Longitudinal Deficiency (Bayne and Klug).

Type	Thumb Anomaly	Carpal Anomaly	Distal Part of Radius	Proximal Part of Radius
N	Absence or hypoplasia	Normal	Normal	Normal
0	Absence or hypoplasia	Absence, hypoplasia, or coalition	Normal	Normal, radioulnar synostosis, or radial head dislocation
1	Absence or hypoplasia	Absence, hypoplasia, or coalition	>2 mm shorter than ulna	Normal, radioulnar synostosis, or radial head dislocation
2	Absence or hypoplasia	Absence, hypoplasia, or coalition	Hypoplasia	Hypoplasia
3	Absence or hypoplasia	Absence, hypoplasia, or coalition	Absence of physis	Variable hypoplasia
4	Absence or hypoplasia	Absence, hypoplasia, or coalition	Absence	Absence

Table 4. Classification of Ulnar Deficiency (Bayne).

Type 0	Deficiencies of the carpus and/or hand only	
Type I	Undersized ulna with both growth centers present	
Type II	Part of the ulna is missing (typically the distal ulna is absent)	
Type III	Absent ulna	
Type IV	Radiohumeral synostosis	
Classification of Ulnar Deficiency According to Abnormality of First Web Space (Cole and Manske)		
Subtype	Grade	Characteristics
A	Normal	Normal first web space and normal thumb
B	Milde	Mild deficiency of first web space and mild thumb hypoplasia with intact opposition and extrinsic tendon function
C	Moderate to severe	Moderate-to-severe deficiency of first web space and similar thumb hypoplasia with malrotation into plane of digits, loss of opposition, and dysfunction of extrinsic tendons
D	Absent	Absence of thumb

Table 5. Radial-ulnar (anterior-posterior) axis formation/differentiation pathology patients treated 2012–2014.

A. Radial Longitudinal deficiency						
Patient	Gender	Type (Bayne)	Associated syndromes	Hand involved	Wrist reconstruction	Thumb reconstruction
1.	M	IV	Holt-Oram	bilateral	Ex fix and radialisation at age 2 and 3	Pollicisation at age 4. Three digit hand
2.	M	IV	TAR	bilateral	Ex fix and 2 nd MTP transfer at age 2 and 3	No thumb hypoplasia
3.	F	IV	no	right	Ex fix and 2 nd MTP transfer at age 2	Second toe transplantation at age 4
4.	F	IV	no	right	Ex fix and centralisation at age 12	No done yet
5.	F	III	no	bilateral	Ex fix and fibula head transplantation at age 10 (left hand)	No done yet
B. Ulnar longitudinal deficiency						
6.	M	IID	no	right	10 month – multiple osteotomies	Sindactily release and spare part surgery at 14 month
C. Ulnar dimelia (mirror hand)						
7.	F		no	right	Extra radial part of ulna removed at age 8 month	'Finger switch procedure' at age 15month

Table 6. Radial club hand treatment results.

Patient	Follow up period (month)	Wrist reconstruction	Time of operation	Wrist		Thumb reconstruction	Grasp kg	Pinch kg	VAS	PEDI	
				flexion	extension					Daily Activities	Mobility
M 4 y	16	2 nd MTP (both)	RH – 4h LH – 3h55`	RH - 30° LH - 35°	RH – 30° LH - 20°	Not needed	RH – 3kg LH – 2kg	RH – 2kg LH -1,5kg	9	56	69
F 5 y	24	2 nd MTP (right)	RH – 3h15`	RH - 10° LH - intact	RH - 90° LH - intact	2 <sup>nd</sup> toe transfer – 4h5`	RH-1,5kg LH - intact	RH – 0,2 LH -intact	10	58	70
M 5 y	48	Radialisation (both)	RH – 1h10` LH – 1h5`	RH - 0° LH - 5°	RH – 25° LH – 10°	Pollicisation (both hands)	Not measured	Not measured	7	51	68

REFERENCES

1. Albee FH. Forming of radius congenitally absent // *Annals of Surgery* 1928;87:105-110
2. Buck-Gramcko D. Congenital Disorders: Radial and Ulnar Club Hand // In : *Hands Surgery* 1st ed. Lippincott Williams&Wilkins;2004:1453-1464
3. Buck-Gramcko D. Radialization as a new treatment for radial club hand. // *The Journal of Hand Surgery* 1985; 1:964-968
4. Chingwundoh J.O.M., Gupta M., Scott W.A. Ulnar dimelia. Is it a true duplication of the ulna? // *The Journal of Hand Surgery* 1997; 22(1): 77-79
5. Gerber EA, Laurell T, Arner M. Epidemiology of Congenital Upper Limb Anomalies in Stockholm, Sweden, 1997 to 2007: Application of the Oberg, Manske, and Tonkin Classification // *the Journal of Hand Surgery [Am]*. 2014; 39(2):237-248
6. James MA, McCarroll HR Jr, Manske PR. The spectrum of radial longitudinal deficiency: a modified classification // *The Journal of Hand Surgery [Am]*. 1999; 24:1145-55
7. Jameel J, Khan AQ, Ahmad S, Abbas M. Ulnar dimelia variant: a case report // *Journal Orthopaedic Traumatology* 2011; 12:163-165
8. Kozin SH. Radial Club hand // <http://emedicine.medscape.com/article/1243998> Oct 24, 2014
9. Kessler I. Centralisation of the radial club hand by gradual distraction // *The Journal of Hand Surgery* 1989; 14(1):37-42
10. Kotwal PP, Varshney MK, Soral A. Comparison of surgical treatment and nonoperative management for radial longitudinal deficiency // *The Journal of Hand Surgery (European Volume)* 2011; 37(2):161-169
11. Kotkansalo T, Elo P, Luukaala T, Vilkki S. Long term effects of toe transfers on the donor feet // *The Journal of Hand Surgery* 2014, 39(9): 966-976
12. Kozin SH. Upper-Extremity Congenital Anomalies // *Journal of Bone Joint Surgery [Am]*. 2003; 85:1564-1576.
13. Oishi SN, Ezaki M. Radial Dysplasia Reconstruction // *Operative Techniques in Orthopaedic Surgery*. Lippincott williams. 2011;51:1376-1379
14. Senes FM, Catena N. Correction of Forearm Deformities in Congenital Ulnar Club Hand: One-Bone Forearm // *The Journal of Hand Surgery* 2012;37(1):159-164
15. Tonkin MA. On the classification of congenital thumb hypoplasia // *The Journal of Hand Surgery* 2014, 39(9) 948-955
16. Takagi T, Seki A, Takayama S. Elbow and forearm reconstruction in patients with ulnar dimelia can improve activities of daily living // *Journal of Shoulder and Elbow Surgery* 2014; 23:68-72
17. Tonkin MA, Tolerton SK, Quick TJ, Harvey I, Lawson RD, Smith NC, Oberg KC. Classification of Congenital Anomalies of the Hand and Upper Limb: Development and Assessment of a New System // *The Journal of Hand Surgery* 2013;38(9): 1845-1853
18. Vilkki SK. Severity grading in radial dysplasia // *The Journal of Hand Surgery* 2014; 39(9) 977-983
19. Vilkki SK. Vascularized Metatarsophalangeal Joint Transfer for Radial Hypoplasia // *Seminars in plastic surgery* 2008; 22(3):195-212

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