Therapy Management of Children with Congenital Anomalies of the Upper Extremity

5

Ginny Gibson

This chapter will first introduce evaluation tools appropriate for children with congenital anomalies of the upper extremity (CAUE). Second general rehabilitation interventions will be described. Third attention will be given to interventions for children with selected CAUE who are often served by occupational or physical therapists.

Evaluation

Reviews, reports, and investigations have identified activities of daily living (ADL) and instrumental activities of daily living (IADL) that are problematic for children with CAUE including styling hair, squeezing toothpaste, completing toilet hygiene, tying shoelaces, closing and opening dressing fasteners, tucking shirts into pants at the waistline, donning socks, cutting and peeling food, and opening containers [1–4]. Educationally related activities that may prove difficult for children with CAUE include writing with a pen or pencil, using a keyboard, carrying books, cutting with scissors, managing a lunch tray, and full participation in playground activities and physical education [4, 5]. Outside of school, children with CAUE have reported difficulty with ball sports, dancing, martial arts, snow or ice sports, water sports, gymnastics, cycling, and playing with construction toys [5]. The evaluation of children with CAUE should consider impairment, activity performance, and activity participation, as there may or may not be a relationship between the three constructs [4, 6, 7].

Four studies have examined the relationship between impairment and body structure with activity performance or participation for children with radial longitudinal deficiency (RLD). Kotwal et al. [6] retrospectively compared children with RLD who underwent centralization or radialization to those who did not. Although the main purpose of the study was to discern if patients benefitted from surgical correction of wrist deformity, the researchers found strong correlations between Prosthetic Upper Extremity Functional Index (PUFI) scores and three measures of body function; including wrist range of motion (ROM) ($r=0.65$–0.81), long finger ROM ($r=0.93$–0.97), and grip strength ($r=0.90$–0.97). Buffart et al. [7] examined relationships between hand function impairment and activity performance. Grip and pinch strength, as well as AROM, were measured for the assisting hand. For children with unilateral involvement the affected hand was measured, and for those with bilateral involvement the more affected hand was measured. All children completed the Assisting Hand Assessment (AHA) and their parents completed the Ease of Performance Scale on the PUFI. Grip strength significantly correlated with activity performance for the AHA ($rp=0.69$, $p=0.002$) and PUFI ($rp=0.52$, $p=0.003$). Pinch strength significantly correlated with activity performance for the AHA ($rp=0.77$, $p=0.001$) only. AROM of the wrist and second digit significantly correlated with activity performance for the AHA ($rp=0.59$, $p=0.006$ and $rp=0.87$, $p=0.001$) and PUFI ($rp=0.71$, $p=0.001$ and $rp=0.59$, $p=0.006$, respectively). Ekblom et al. [8] found a relationship between outcomes on the AHA and total ROM of digits ($p=0.042$), and self-experienced time of performance on the Children's Hand-use Experience Questionnaire (CHEQ) and total active motion of the wrist ($p=0.043$). There was no relationship between the degree of radial deviation and outcomes of the Box and Block Test, AHA, or CHEQ. The aforementioned studies included children.

Holtslag et al. [4] investigated the functional implications of RLD for 17 adults who previously underwent surgical or conservative treatment. Measurements included grip and pinch strength, ROM, and hand function during standardized ADL using the Sequential Occupational Dexterity Assessment (SODA). Participation in activity was quantified using the

G. Gibson, O.T.D., O.T.R./L., C.H.T. (✉)
Pediatric Rehabilitation, UCSF Benioff Children's Hospital Oakland, 747 52nd Street, Oakland, CA 94609, USA

Department of Occupational Therapy, Samuel Merritt University, 450 30th Street, Oakland, CA 94609, USA
e-mail: ggibson@samuelmerritt.edu

D.R. Laub Jr. (ed.), *Congenital Anomalies of the Upper Extremity: Etiology and Management*,
DOI 10.1007/978-1-4899-7504-1_5,

Table 5.1 Impairment-based measurement tools or techniques

Impairment	Measurement	Impairment	Measurement
Movement restriction	Goniometer	Impaired sensation/nerve injury	Monofilaments
	Inclinometer		Two-point discrimination
	Wire tracing		Stereognosis
	Tape measure		Moberg pickup test
	Pollexograph		Ten test
			Ninhydrin sweat test
Weakness	Dynamometer	Prehension	Box and blocks
	Quantified muscle testing (QMT)		Nine hole peg test
	Manual muscle testing (MMT)		Functional dexterity test
Edema	Volumetry		Test of in-hand manipulation
	Tape measure		
Pain	Visual analog scale		
	Wong-Baker faces		
	Face, legs, activity, cry, consolability scale		

Table 5.2 Impairment ratings

		Normative data				
Impairment	Tool	First author and date	Citation	Measure	*N*	Age (years)
Movement restriction	Goniometer	Soucie (2011)	[9]	Shoulder, elbow, and forearm PROM	200	2–19
	Goniometer	Barad (2013)	[10]	Elbow PROM	1,361	1–16
	Pollexograph	de Kraker (2009)	[11]	Thumb abduction	100	4–12
	Distance measurement					
Weakness	Grippit	Häger-Ross (2002)	[12]	Grip strength	530	4–16
	Jamar dynamometer	Bowman (1984)	[13]	Grip strength	153	6–9
	Jamar dynamometer	Fullwood (1986)	[14]	Grip strength	214	5–12
	Jamar dynamometer	De Smet (2001)	[15]	Grip strength	487	5–15
	Jamar dynamometer	Holm (2008)	[16]	Grip strength	376	7–12
	Jamar dynamometer	Ploegmakers (2013)	[17]	Grip strength	2,241	4–15
	Jamar dynamometer	Mathiowetz (1986)	[18]	Grip strength	571	6–19
	B & L pinch gauge			Pinch strength		
	Preston pinch gauge	Lee-Valkov (2003)	[19]	Pinch strength	17	3–5
	Jamar dynamometer			Grip strength		
	Preston pinch gauge	Ager (1984)	[20]	Pinch strength	474	2–13
	Jamar dynamometer			Grip strength		
	B & L pinch gauge	Surrey et al. (2001)	[21]	Pinch strength	414	5–12
	Preston pinch gauge	De Smet (2006)	[22]	Pinch strength	262	5–12
Impaired sensation	Two-point discrimination	Cope (1992)	[23]	Discriminative touch	112	2–13
Prehension	Box and block test of manual dexterity	Mathiowetz (1985)	[24]	Manual dexterity	471	6–19
	Functional dexterity test	Gogola (2013)	[25]	Manual dexterity	175	3–17
	Nine hole Peg test	Smith (2000)	[26]	Manual dexterity	826	5–10
		Poole (2005)	[27]	Manual dexterity	409	4–19
	Purdue Pegboard	Wilson (1982)	[28]	Manual dexterity	206	2.6–5.1

Normative studies in typically developing children

Impact on Participation and Autonomy (IPA) questionnaire. Researchers found a positive correlation ($r=0.56$, $p=0.02$) between digital ROM and SODA outcomes, but no other relationship between body function and hand function or participation. Table 5.1 includes tests and measures of impairment, likely to be used when providing habilitative and rehabilitative services to children with CAUE, while Table 5.2 includes tests for which normative information is available.

Table 5.3 Performance-based assessment

Assessment tool	Target populations	Target age (years)	Studies describing psychometrics		
			Validity	Reliability	Responsiveness
ABILHAND-Kids	Children with cerebral palsy	6–15	[33]	[33, 35]	–
AHA	Children with typical function in one hand only	1.6–12.8	[36]	[33, 37, 38]	[37]
CHEQ	Children with typical function in one hand only	6–18	[39]	–	–
PedsQL	Children with acute or chronic illness	2–18	[40]	[40]	[41]
PEDI	Children with physical disability	6 months–7.5	[42–44]	[45]	–
PODCI	Children with orthopedic conditions	0–18	[46, 47]	[46]	[48, 49]
PUFI	Children who use an upper extremity prosthesis	3–18	[33, 50]	[33, 51]	–
UBET	Children with transverse reduction deficiency	2–21	[33]	[33, 52]	–

AHA Assisting Hand Assessment, *CHEQ* Children's Hand-use Experience Questionnaire, *PedsQL* Pediatric Quality of Life Inventory, *PEDI* Pediatric Evaluation of Disability Inventory, *PODCI* Pediatric Outcomes Data Collection Instrument, *PUFI* Prosthetic Upper Extremity Functional Index, *UBET* Unilateral Below Elbow Test

Skerik et al. [29] described a standardized process of assessment for all children with CAUE including analysis of available patterns of pinch and grip, observation of preferred patterns of usage, and measurement of ROM, pinch strength, and hand size. To measure outcomes following index finger pollicization, Percival et al. [30] developed a battery of seven tests for which a maximum score is 22. Included in this battery is a measure or observation of tip pinch and pulp pinch strength; opposition of the thumb to the middle, ring and small finger; grasping of two balls of different size, active movement of the thumb at three joints; two-point discrimination; and cosmesis (length and position of the thumb). Scores are characterized as excellent (>20) good (16–19), fair (12–15), or poor (<12). Ho and Clarke [31] conducted a systematic review of studies published between 1966 and 2003 aimed at evaluating outcomes following pollicization of the index finger or centralization for radial longitudinal deficiently. Of the ten studies reviewed, six attempted to measure ADL or functional use of the hand, but only one did so using a standardized instrument.

Since Ho and Clarke's review [31], other outcome studies have been published in which standardized assessment tools were employed. Buffart et al. [32] set out to identify appropriate assessment tools for use with children with transverse or longitudinal reduction deficiency using as criteria inclusion of bimanual tasks, measures of quality of movement, and appealing tasks. These researchers recommended the AHA, Unilateral Below Elbow Test (UBET), ABILHAND-Kids, and PUFI. In a follow-up study [33] the AHA, UBET, ABILHAND-Kids, and PUFI were administered to 20 children with RLD, aged 4–12 years. The AHA and PUFI were deemed most valid for children with RLD, due to the relationships found with type of RLD ($r=-0.82$ and -0.64, respectively), functional hand grips ($r=0.58$ and 0.46, respectively), and the therapist's global assessment of hand function ($r=0.85$ and 0.63, respectively). Kaplan and Jones [34] used the Pediatric Outcomes Data Collection Instrument (PODCI) to determine outcomes following microsurgical toe transfers for thumb reconstruction. Table 5.3 presents performance-based assessments specifically designed for children with CAUE, children with normal use of one hand only, or children with disability but no specific diagnostic population.

Assessments that attempt to measure satisfaction with or perceptions of activity performance and participation should include the child with CAUE, but in some cases the caregiver may need to serve as proxy. Researchers have studied the extent to which parents and children agree on satisfaction with or perception of activity performance and participation. Netscher et al. [53] examined ability to participate in activities following index finger pollicization. In addition to measuring impairment level and ability to participate in simulated tasks reflecting participation in a larger activity, researchers administered a non-validated novel questionnaire to nine children and their parents to determine perceptions of appearance, social participation, and performance skills. The mean score for children was 22, with 12 being the best score and 60 the worst. Although parents tended to assess their children's skills as slightly better than the children did of themselves, there was no statistically significant difference between parents' and children's scores, suggesting that parents may serve well as proxy. Ardon et al. [54] found similar results when parents and their children with CAUE separately completed the Pediatric Quality of Life Inventory (PedsQL). No statistically significant differences were observed for total score and the five domains (physical health, emotional functions, social functioning, school functioning, psychosocial health). The researchers noted analysis of individual scores showed children and parents tended to disagree and the variables that influenced disagreement included number of affected digits and bilateral involvement [54]. Similarly, in a large multi-center study significant differences were found between parents and their children with congenital below elbow deficiency (CBED) for upper

extremity physical function ($p<0.001$), pain/comfort ($p<0.05$), and social functioning ($p<0.001$) using the PODCI and PedsQL [55]. In summary, use of a parent as proxy should be limited; effort to elicit children's participation is desirable.

Interventions to Address Impairments

Current estimates of the rate of congenital upper limb differences include 1 in 506 live births [56], 5.25 in 10,000 live births [57], and 21.5 per 10,000 live births [58]. In two reports of the incidence of all congenital limb reductions, 75–81 % involved the upper extremities [59, 60]. Within these estimates, not all children with CAUE will require surgical intervention and subsequent rehabilitation. When indicated, rehabilitation efforts may initially emphasize interventions to address impairment with simultaneous or subsequent attention to participation in activity. The studies presented in the following sections are not specific to children.

Edema

Edema management is often addressed via rest, ice, compression, and elevation. Postoperative dressings and casting provide rest and compression yet preclude icing. Chronic edema that persists after removal of postoperative immobilization may be treated with gentle compression. Younger children may not be amenable to elevation and the efficacy of elevation following hand surgery is unclear. In two prospective and randomized comparison trials, no statistically significant differences were noted in those who the limb and those who did not for adults undergoing Dupuytren's release [61] or carpal tunnel decompression [62]. Gentle compression may be achieved with self-adhering wrap [63]; however, only one case report of an adult with burn injury could be located to support its use [64].

Scar

In addition to being cosmetically unappealing, postoperative scar may lead to motion restriction, pain, and puritis. These impairments may in turn reduce function and participation in activities. Intervention should first concentrate on prevention of hypertrophic scars, but when hypertrophic scars are present, efforts should be made to reduce the extent of the existing scar. Scars from surgical incisions may respond well to treatment including massage, pressure, topical application of a gel product, and reduction of tension on scar.

Massage

Shin and Bordeaux [65] conducted a systematic review of studies investigating the effectiveness of scar massage regimes for scars due to burn and trauma, and included four randomized controlled studies, three prospective controlled studies, one prospective study, and two case reports. Across ten reports, the total number of subjects was 220 with 144 receiving scar massage. The standardized outcome measures included the Observer Scar Assessment Scale and the Vancouver Scar Scale (VSS), as well as subjective assessments of scar thickness, perfusion, color, pain, and itching. For patients who had surgical scars and received massage, 90 % improved. Foo and Tristani-Firouzi [66] recommend that postsurgical scar massage commence during the proliferative phase, 2–3 times per day, for 3–5 min, for 3–4 months.

Pressure

Pressure application may be applied using self-adherent wraps, neoprene splints, tubular elastic, and custom fit pressure garments. Pressure inhibits fibroblastic activity [66], via ischemia and hypoxia resulting in degeneration of fibroblasts and slowed synthesis of collagen [67]. Despite a long history of inclusion of pressure in the treatment of scar, definitive evidence regarding its efficacy is lacking.

In a meta-analysis of six published randomized controlled trials and one unpublished trial examining the benefit of pressure therapy for burn scar, researchers found no difference between scars treated with pressure therapy and controls [68]. More recently, a randomized controlled study of treatment of burn scar demonstrated significant improvement on the VSS using pressure therapy alone ($p<0.001$), but also found significant improvement with combined pressure therapy and application of silicone gel sheeting ($p=0.001$), and combined pressure therapy and silicone spray ($p<0.001$). Patients with two similar scars from split-thickness grafts were randomized into either a silicone gel sheeting group or silicone spray group, but all used pressure therapy. Differences between the groups were not significant [69]. Widgerow [83] suggests pressure garments are more appropriate for widespread scar seen in burn injury; however, in maintenance of tape or silicone gel sheeting on the hand of a young child can be challenging. Use of a pressure garment may discourage self-removal of treatment modalities held in place by the garment, including the garment itself. In a laboratory study of fibroblastic activity under pressure, researchers showed pressure application may be applied at higher levels over shorter periods of time or at lower levels for longer periods of time to reduce fibroblastic proliferation [66].

If using self-adherent wrap, care in wrapping and maintained supervision are indicated to avoid a tourniquet-like effect due to lifting, slippage, and rolling [72]. Use of neoprene

patches or orthoses for at least 8 h per day was retrospectively studied in a small population of children and young adults with burn scar ($n=8$ participants, 12 scars). Duration of treatment ranged from 1 to 11 months. Scars were evaluated pre and post treatment and differences for mean VSS was significantly lower after treatment ($p=0.0001$). This study is useful to therapists working with children because neoprene splints are often used long term across several diagnostic groups for limb positioning and so could also serve to manage scar [73].

Silicone

Silicone gel may serve to prevent hypertrophic scars and improve characteristics of existing hypertrophic scar. In a narrative review of eight RCTs and an analysis of 27 trials, the International Advisory Panel on Scar Management concluded that use of silicone gel sheeting is "safe and effective"; however, the panel distinguished adhesive silicone gel sheeting from other adhesive gels, liquid silicone, and nonadhesive silicone gel sheeting [74]. O'Brien and Pandit [75] conducted a meta-analysis to determine the effectiveness of silicone dressings to prevent hypertrophic or keloid scarring in people with newly healed wounds and to treat established keloid or hypertrophic scars. The study included randomized or quasi-randomized controlled trials, and controlled clinical trials comparing silicone dressings to other nonsurgical treatment, no treatment or placebo. Included trials compared adhesive silicone dressings with control; non-silicone dressings; silicone gel plates with added vitamin E; laser therapy; triamcinolone acetonide injection, and nonadhesive silicone dressings. Scar quality was determined by blood flow, color change, hyperpigmentation, thickness, and shape. Studies that set out to determine effectiveness of silicone to treat existing scars measured change in scar size and did so using a ruler, taking an impression, or via ultrasound. Across 15 studies, 615 people between 2 and 81 years-of-age were included. Compared with no treatment silicone reduced the incidence of hypertrophic scar (RR 0.46, 95 % CI 0.21–0.98). For established keloid and hypertrophic scar SD significantly reduced scar thickness (RR −1.99, 95 % CI −2.13 to −1.85) and improved color (RR 3.05, 95 % CI 1.57–5.96). Silicone dressings produced superior results compared to controls in two trials, no difference was found in two trials, and the control group fared better in one trial. This study included clinical trials of varied rigor and most were subject to bias thus there is weak evidence for use of silicone dressing to prevent or improve scars. An update to this review was published in 2013; five new studies were included but the same conclusion was offered [76].

The proposed mechanism of action of silicone gel is thought to be hydration and occlusion [77], though non-silicone gels may be equally effective as silicone. In a prospective, randomized study patients ($n=24$) with existing hypertrophic or keloid scars ($n=41$) present for longer than 3 months, including incisional scars, were randomly assigned to one of three groups: treatment with silicone gel ($n=16$ scars), treatment with non-silicone gel ($n=14$ scars), or control ($n=11$ scars). Treatment was applied 24 h per day for 4.5 months. No statistically significant differences were found between SD and NSD groups for color, size, induration, and symptoms, although significant differences were noted when SD and NSD were compared to controls for color, size, induration, and scar pliability [78].

Tape

Tension on scar is believed to stimulate collagen production due to mechanosensitive fibroblasts [71, 79, 80]. Tape applied to scars may reduce tension and prove effective in preventing hypertrophic scar [81, 82]. Porous tape should be applied longitudinal to and directly over the scar to adequately provide support and reduce tension [83]. When scars cross joints, use of an orthosis may help to reduce tension on scar.

Motion Restriction

Clinicians utilize AROM, active assisted ROM, passive range of motion (PROM), joint mobilization and orthoses to achieve greater range of movement. Michlovitz et al. [84] conducted a systematic review of interventions to promote joint motion in the upper extremity. The review included 26 studies that examined interventions in adults, but excluded children and congenital hand differences. In their summary, the researchers noted moderate evidence for the use of orthoses or casts and passive exercise to increase ROM after joint trauma or immobilization. Following this study, Glasgow et al. [85] published a narrative review to develop a set of recommendations for mobilizing the stiff hand. After a review of 29 studies of varying levels, these authors recommended active and active assisted exercise during all stages of tissue healing, passive exercise during the proliferative and remodeling phases, and joint mobilization during the remodeling phase. Orthoses for management of stiffness via mobilization were recommended during the proliferative and remodeling phases.

When the purpose of an orthosis is to increase motion, orthosis prescription must consider tissue compliance and the length of time the restriction has been present. Therapists must decide on orthosis type (including no orthosis), wear time (hours per day and duration), and the magnitude of force to apply. Flowers [86] offered a hierarchy for decision-making when treating stiff joints using a modified Week's test [87]. After pre-conditioning, those whose PROM measures change by 20° may not need a splint; by 15° may require a static splint with no overpressure; by 10° may require a dynamic splint; and by 5° or less may required a static progressive splint with overpressure. This decision-making process may prove useful with older children; but may not be feasible with infants and toddlers due to required exposure to thermotherapy.

Consensus on wear time of an orthosis to resolve motion restrictions is lacking, although many studies provide guidance. Flowers and LaStayo [88] executed a study to determine if duration of orthosis use impacted outcomes for stiff joints. Patients ($n=15$) with 20 PIP flexion contractures between 15° and 60° were randomly assigned to continuous casting for 6 days then 3 days or 3 days then 6 days. There was a statistically significant difference ($p<0.005$) in gains made with 6 days of wear achieving a mean increase of 5.3° and 3 days of wear achieving 3°. Glascow et al. [89] prospectively investigated optimal hours of daily orthoses wear in 43 subjects with joint restrictions in the hand following trauma. Subjects with similar levels of stiffness—as determined via torque range of motion (TROM)—were randomly allocated to a <6 h or 6–12 h per day group. There was a statistically significant difference between the groups, with better TROM observed in the 6- to 12-h group. It is not clear if increasing time more than 12 h provides greater benefit. In a follow-up randomized study of 22 patients with PIP joint flexion contractures, no significant differences were found for PROM, AROM, or TROM between 6–12 h of wear and 12–16 h of orthosis wear after 8 weeks of treatment [90].

Interventions to Address Activity Performance and Participation

Assuming a child with CAUE is otherwise typically developing, interventions to improve activity performance or participation may occur immediately following surgery or intermittently—when the child encounters specific problems with activity performance or participation. Following surgery, impairment-based may be emphasized concurrently with interventions to promote activity performance and participation via activity modification or introduction to assistive devices [4]. In a qualitative study investigating perceptions of children 8–20 years of age with unilateral CBED, participants described their own activity performance and participation and generally reported no limitations. Further, these children reported similar levels of participation as peers without CBED. The researchers suggested, for children in this study, perceptions of activity participation might have been limited to actual chosen activities rather than potential chosen activities (activities that may have been chosen if participants had two hands) [5]. In a descriptive study of eight people with ulnar longitudinal deficiency (ULD), age 3–41, adult patients reported no difficulty with self-dressing, washing, toileting, eating, closing and opening dressing fasteners, managing the telephone, typing, or opening containers with screw on caps and parents of children with ULD reported no difficulty with bimanual self-care, play, or school related activity [91].

Health professionals should recognize there are multiple strategies to manage limitations in activity performance and participation that may be acceptable to the child with CAUE including using other body parts (Fig. 5.1), activity modification, choosing varying levels of participation, receiving assistance from another, using assistive devices, and prosthetic wear [5]. In the study by de Jong et al. [5] health professionals were less apt to recognize as many strategies as did children and their parents, and identified assistive devices and prosthetics more frequently as potential solutions for success in activity performance and increased participation.

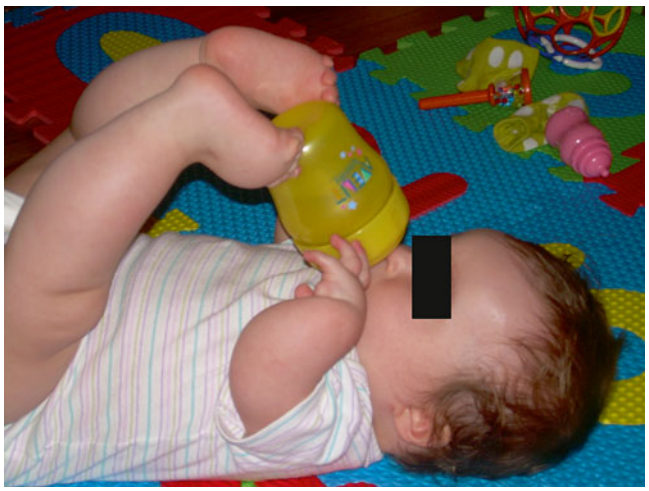

Fig. 5.1 This child with thrompocytopenia absent radii has self-identified strategies for participation in activities

Diagnosis-Specific Intervention

Camptodactyly

Range of Motion Exercise

While orthotic management and surgery are intervention options for camptodactyly [92], ROM exercises may prove beneficial especially for children with an infantile onset of deformity. Rhee et al. [93] retrospectively evaluated the effectiveness of passive stretching to correct flexion deformities in children younger than 3 years with camptodactyly. Records of children with simple camptodactyly who had not received surgery or intervention with an orthosis were included, but those with flexion contractures of less than 10° were excluded. Parents were taught a PROM technique, to be implemented at home, requiring the PIP joint be extended with the wrist and metacarpophalangeal (MCP) joint in extension. Instructions were to complete gentle PROM, while the child was sleeping, 20 or more times per day with a hold time of 5 min. Exercise frequency was reduced to five or ten times per day when near full extension was achieved. Duration of intervention was individualized and poorly defined. The intervention could be realistically applied; however, the burden of applying PROM

only when children are sleeping could compromise adherences rates. Pre- and post-intervention measurements, recorded by the same physician, were compared. Across groups, 13 males and 9 females with a mean age of 12 months (range 3–36 months) were included in the study. Digits were further classified into mild deformity (<30°, $n=12$ digits), moderate deformity (30–60°, $n=36$), and severe deformity (>60°, $n=13$) as per goniometric measures. Groups were expected to be different with regard to extent of deformity but no analysis was performed to assure they were similar for age, sex, and dominance. Final PROM for PIP extension was compared to initial measures. Mean change in PROM were as follows: −20° to −1° for the mild group, −39° to −12° for the moderate group, and −75° to −28° for the severe group. Differences from pretest to posttest were significant for all groups: mild ($p<0.001$), moderate ($p<0.001$), and severe ($p<0.001$). Mean time from start to end of intervention (either correction or cessation of change) for the mild group was 5 months, moderate group was 10 months, and severe group 13 months. Researchers found a relationship between degree of flexion contracture at the start of intervention and final measure. No relationship was found between initial flexion contracture, handedness, digit involvement, and number of digits or hands involved. Differences between pretest and posttest AROM values were statistically significant. No statistical analysis was performed to determine clinical significance, however all but two children (in the moderate group) improved and gains were maintained during a prolonged follow-up period (mean of 26 months, range of 12–47 months). The researchers concluded children under three who have camptodactyly should be treated with PROM only and orthoses are not necessary; however, this statement is unfounded since no comparison was made between PROM and use of an orthosis. The researchers recognized the weaknesses of the study including use of retrospective design and absence of a control group. The outcomes cannot be applied to all children with camptodactyly since only children under the age of three with simple syndactyly were studied, and children with syndromic or adolescent onset camptodactyly were not included [93].

Orthotics

In a descriptive case series, Hori et al. [94] evaluated the effectiveness of dynamic splinting on increasing digital extension in 24 (34 fingers) children with camptodactyly. A Capener type coil spring was applied initially for 24 h per day and then only 8 h per day during a maintenance period. Duration of treatment was individualized and not described. Measurement technique was unclear in 10 patients but an explicit statement regarding measurement was provided for 14 patients (21 fingers). The researchers noted "almost full correction" [94, p. 1062] in 14 patients (20 fingers). Eight patients (nine fingers) improved, three fingers were not improved, and two patients (two fingers) worsened. Of the 14 patients (21 fingers) measured, mean flexion contracture before and after intervention was 40° and 10°, respectively. Reoccurrence was noted in one patient. This study lacked a control group, randomization, and blinding. No statistical analysis was undertaken thus limiting generalizations to the larger population. Significant bias is likely since for some patients, AROM may have been determined by visual observation alone.

Miura et al. [95] also examined the effectiveness of dynamic splinting on increasing digital extension but did so prospectively and included a larger sample than Hori et al. [94]. The study included children ($n=142$) with non-traumatic flexion deformities. Of these, 62 had small finger involvement, 16 had small finger plus one or more other finger involvement, 41 had other finger involvement (not small finger), and 23 had syndromic camptodactyly. A dynamic orthosis (Capener type coil spring) was applied to children with contracture of the small finger only for 24 h per day, although only 12 h per day for children under 7 years of age. During a maintenance period wear time was reduced. Outcomes were dichotomized into *failed to respond* or *responded* to treatment. Of 142 patients, only five failed to respond to treatment. Reoccurrence was observed in two patients. From this study alone no definitive statements can be made regarding treatment of children with camptodactyly using orthoses; however, given the number of patients who made gains low-level evidence if offered [95]. Figure 5.2 depicts a serial static orthosis used to correct improve joint motion in camptodactyly.

Orthotics and PROM

Benson et al. [96] retrospectively evaluated the effectiveness of orthoses and PROM to conservatively treat camptodactyly across three subtypes involving the PIP joint. In this case series, in which only descriptive analysis was performed,

Fig. 5.2 Orthosis for camptodactyly involving multiple digits

researchers treated contracted digits of 18 patients (50 PIP joints) to promote PIP extension. Wear time for the orthosis ranged from 15–18 h per day for infants and 10–12 h per days for older children who were not inclined to sleep during the daytime. Parents performed PROM daily prior to application of the orthosis, although duration of treatment was individualized and poorly defined. Using goniometry, the same rater measured PROM before and after the intervention period. For analysis, children were assigned to one of three groups, including: (1) infantile camptodactyly between the age of 0.3–2.3 years ($n=13$ patients, 24 digits); (2) adolescent camptodactyly between the age of 14.5–17.0 years ($n=4$ patients, 5 digits); and (3) syndromic camptodactyly between the ages of 0.1–13.4 years ($n=5$ patients, 30 digits). Full passive extension was achieved in 18 of 24 PIP joints for children with infantile camptodactyly. The group mean at start and end of treatment was −22.9° and end −4.3°, respectively. For children with adolescent onset of camptodactyly, only one (1 PIP joint) underwent a full program of orthosis wear and achieved full extension. Two others (2 PIP joints) elected surgery and worsened. The fourth patient (2 PIP joints) abandoned orthosis wear after 1 month and worsened. The group mean at start and end of treatment was −29.0° and −32.0°, respectively. In the syndromic group, four patients (24 PIP joints) were treated with an orthosis and demonstrated a group mean at the start and end of treatment of −23.0° and −1.0°, respectively. Two patients elected surgery and gained motion; one achieved full extension in two of two PIP joints and the other achieved and average of 41° of improvement across four digits. This study suggests conservative management with an orthosis may be prudent prior to electing surgery and perhaps more so with patients who present with infantile camptodactyly; however, in the absence of a control group, randomization, blinding, long-term follow-up, and inferential statistical analysis the outcomes are inconclusive [96].

Hypoplasia of the Thumb

Therapy interventions for children with thumb hypoplasia will vary greatly depending upon the severity of involvement and surgical management. This section will include interventions for children undergoing surgical procedures for Grade IIIA hypoplasia including web deepening, stabilization of the MCP joint and tendon transfers, and those for Grade IIIB, IV, and V including pollicization or free toe transfer.

Range of Motion Exercise

First Web Space Deepening, MCP Stabilization, Opponensplasty

At 6 weeks following abductor digiti minimi opponensplasty, supervised AROM and light activity is commenced [97, 98], with emphasis on opposition and palmer abduction, and PROM may commence 8 weeks following surgery [98] as well as resistive pinching [97]. Budding taping of the index finger to the middle finger may help to promote opposition of the index to the thumb by restricting lateral prehension between the index and middle finger.

Pollicization or Free Toe Transfer

For pollicization and free toe transfer, Egerszegi [99] recommends initiation of AROM at 3–4 weeks and PROM 1–2 weeks later; however, Goldfarb et al. [98] suggest PROM not begin until 8 weeks following surgery. Buddy taping of the middle finger to the ring finger may encourage opposition of the pollicized index finger to the middle finger by restricting lateral prehension between the middle and ring fingers.

Orthotics

First Web Space Deepening, MCP Stabilization, Opponensplasty

Following abductor digiti minimi opponensplasty, the hand and wrist should be completely immobilized for 4–6 weeks, after which an orthosis is fabricated to maintain a wide, open web space in opposition and palmer abduction [97, 98]. de Roode et al. [97] specifically recommend a neoprene orthosis. Regardless of type, the orthosis should be worn continuously until the eighth postoperative week [97, 98], and removed only for washing, and supervised activity and exercise. Goldfarb et al. [98] recommend discontinuing all orthoses 12 weeks following surgery, whereas de Roode et al. [97] recommend weaning of the splint to night-time wear only but do not indicate when or if night-time splinting should be discontinued. Goldfarb et al. [98] recommend similar immobilization regardless of opponensplasty technique; however, Kozin and Ezaki [100] recommend a long arm thumb spica cast with the elbow 90° flexion for only 2–3 weeks after flexor digitorum superficialis opponensplasty. These authors did not indicate need for a thermoplastic splint following cast removal.

Pollicization or Free Toe Transfer

Regarding pollicization, Egerszegi [99] recommends continuous immobilization for 3–4 weeks with a thermoplastic orthosis replacing the postsurgical splint and worn continuously for an additional week followed by an additional 6 weeks of orthotic use at night and during vigorous activity. In the case of free toe transfer, a similar program of orthosis wear is indicated except evidence of bony union signifies discontinuance of fulltime wear of an orthosis and transition to night-time wear. Goldfarb et al. [98] recommend an orthosis that places the thumb in opposition and palmar abduction.

Scar Management

Scar management may be initiated as early as 3–4 weeks with scar massage. A pressure garment with silicone sewn

into the garment may prove useful for young children; whereas, gel and elastomer could be held in place with self-adherent elastic wrap for older children.

Radial Longitudinal Deficiency

Range of Motion Exercise

Children with RLD may have limitations in elbow motion in addition to wrist deformity [1, 101]. Brooks [2] recommends active and passive elbow ROM for 5–10 min, five times per day. In a series of 27 children with RLD and restriction in elbow flexion, Lamb [1] observed an increase in active elbow flexion for 20 children when an orthosis was applied to the wrist. Restricting wrist motion may facilitate greater elbow motion by preventing a functional pattern of wrist radial deviation to bring the hand toward the trunk and face. Initially, PROM alone may be indicated to preserve tissue length when the wrist can easily be brought into a neutral position. Bednar et al. [102] recommend passive ulnar deviation for 5–10 min, 4–5 times per day. If not passively correctable to neutral, the addition of orthotic management should be considered. Damore et al. [103] recommend passive ROM only until 3 months of age at which time a night-time only orthosis is introduced.

Following centralization procedures, Goldfarb et al. [98] recommend digital ROM begin immediately, and supervised light active use of the hand out of the orthosis by 6 weeks. Further wrist ROM (excluding passive radial deviation) may begin 6 months following surgery.

Orthotics

Conservative management of RLD includes use of casts or orthoses to preserve (Fig. 5.3) or increase tissue length and increase function [1, 104, 105]. The required duration of orthosis wear to achieve or approximate passive correction will depend upon the degree of deformity and the load required to bring the wrist toward neutral; however, this may need to be balanced by time out of the orthosis for play exploration and maintenance of skin integrity. Use of an orthosis may continue until skeletal maturity [2]. Fuller [105] recommends the orthosis be applied radially but cover 80 % of both the volar and dorsal forearm.

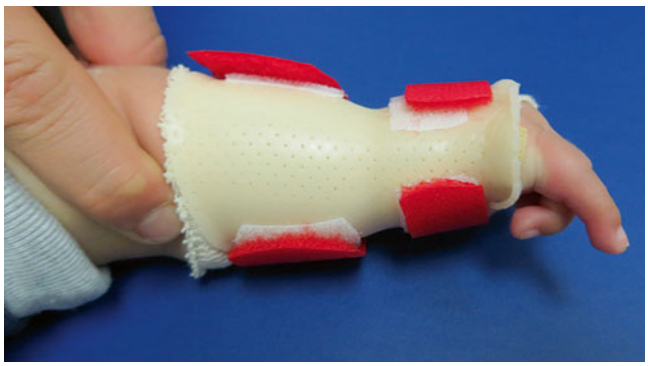

Fig. 5.3 Orthosis for RLD with thumb aplasia

Children with RLD who undergo centralization of the ulna or other soft tissue procedures will require prolonged use of an orthosis pre- and postoperatively [102, 106, 107]. Many authors recommend commencing with orthosis use or serial casting soon after birth and continuing until surgery [102–104, 106]. Kotwal et al. [6] proposed aggressive preoperative use of an orthosis minimizes the amount of tissue disruption and subsequent fibrosis that would otherwise contribute to further deforming forces on the wrist. Following centralization, radialization, or other soft tissue procedure to better align the wrist, fulltime orthosis wear followed by night-time only wear may be indicated. Following centralization and 6–8 weeks of pinning and postoperative splinting, Damore et al. [103] employs fulltime orthosis use followed by weaning toward night-time wear until skeletal maturity. Goldfarb et al. [98] recommend discontinuing use of the orthosis during the day by 6 months but continuing night-time wear until skeletal maturity. For both centralization and radialization, and after 8–12 weeks of internal fixation and splinting, Kotwal et al. [6] introduced fulltime use of an orthosis for 1 year followed by intermittent daytime use for an additional 1–2 years. No mention was made of night-time use during this latter period [6].

Kennedy [108] reported outcomes after applying orthoses to correct excessive radial deviation and minimize soft tissue reconstruction during corrective surgery, or to maintain or improve correction postoperatively. In this case series, children with RLD using an orthosis were treated preoperatively ($n=5$) or postoperatively ($n=4$). In the preoperative group, there were four males and one female with ages ranging from 3 weeks to 5 years. In the postoperative group, there were three males and one female with ages ranging from 2 years to 9 years. Each child received a custom fabricated neoprene orthosis with thermoplastic reinforcement to centrally align the hand to the carpus. Children wore the orthosis fulltime in all environments typical for the child. Duration of treatment for the preoperative group ranged from 3 weeks to 6 months to achieve correction, whereas duration of treatment for the postoperative group ranged from 6 weeks to 2 years to achieve correction. Wrist alignment was the desired outcome but the measurement technique was not described. For the preoperative group, all children obtained a neutral wrist with four children achieving 90° and one 45° of improvement. For the postoperative group mean correction of residual deformity in three children was 30°. Correction was maintained in the fourth child. The author reported subjective observations of improved activity participation with the orthosis including use of cutlery and tying shoelaces. This study lacked a control group, randomization, blinding, use of objective repeatable measures, and statistical analysis, but thoroughly describes a splint and provides descriptive outcomes for a small group of children with RLD [108].

Assistive Technology

Holtslag et al. [4] examined participation levels among adults with mild and severe RLD using the Participation and Autonomy questionnaire (IPA). No significant differences were noted between the groups, and both groups exhibited good levels of participation (median IPA score) 2.4 (a score of zero is very good and a score of four is poor). Some participants in this study indicated a need for activity modification or assistive device to perform fastening of buttons, squeezing a tube of toothpaste, carrying heavy objects, and cutting food. In a series of 117 patients with RLD, Lamb [1] noted no functional impairment for children with unilateral RLD, but for those with bilateral impairment fastening buttons, cutting meat, combing hair, and putting on socks proved difficult. Buffart et al. [7] also identified specific activities found to be difficult for children with RLD including fastening buttons, spreading jam, donning gloves, and cutting firm textured foods. These are important activities to practice with children and, perhaps, introduce assistive devices.

Syndactyly

Complications following syndactyly release include web creep, rotational and angular deformities, and limitations in AROM [109–113] for which ROM, application of an orthosis, and scar management may be indicated [113].

Range of Motion Exercise

Fuller [105] recommends parents be taught PROM. Extension deficits (flexion contractures) can be managed using an orthosis, while limitations in active flexion might be better managed with combined PROM and AROM during the day.

Orthotics

Goldfarb et al. [113] noted patterns of deformity following syndactyly release, for children with complex syndactyly not related to a syndrome or other CAUE, including a trend for the released digit to rotate away from and deviate toward the previously adjoined digit. After the postoperative dressings and splint have been discontinued, a thermoplastic splint may be indicated to maintain the MCP joint(s) in abduction, to correct an extension deficit, or to align the digits along the horizontal and frontal planes. Fuller [105] recommended a static forearm based orthosis with elevation of the material between adjacent digits and individual finger straps, whereas Moran and Tomhave [114] recommend a hand-based orthosis with individual finger straps.

Scar Management

Scar management options may be narrowed since children with syndactyly often undergo release during the infant or toddler years, and so choice of modality must include products that are less likely to pose a choking hazard. For this reason, a pressure garment with silicone sewn into the garment may prove useful for young children; whereas, gel or elastomer (Fig. 5.4) could be held in place with self-adherent elastic wrap for older children.

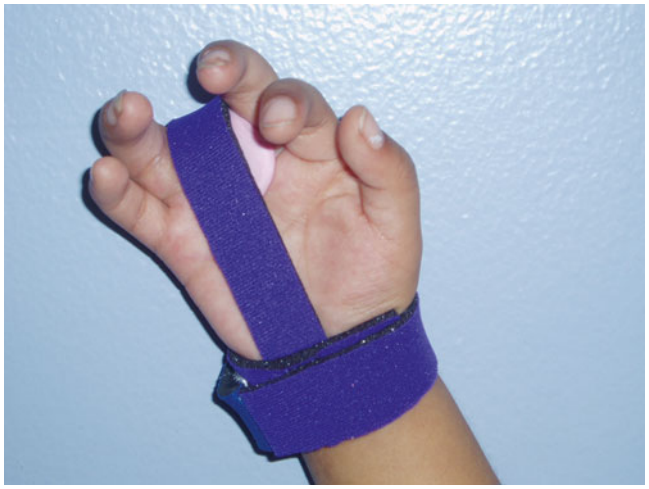

Fig. 5.4 Scar pad for web creep following syndactyly release

Trigger Thumb

Baek and Lee [115] conducted a prospective observational study of 71 trigger thumbs in 53 children whose mean age when diagnosed was 2 years with a mean flexion contracture of 26°. These children were followed for 49 months. Forty-five of 71 thumbs (63 %) spontaneous resolved. For this reason, children with trigger thumb are often observed or offered conservative treatment, including ROM and application of an orthosis.

Range of Motion Exercise

Two groups of researchers [116, 117] favor PROM over use of an orthosis for conservative treatment of trigger thumb. In a prospective, case series, Wantabe et al. [116] described 58 thumbs in 46 children treated with daily passive extension exercises only. Thumbs were identified as: Stage 0: No trigger or flexion posture; Stage 1: Locking, active movement with triggering; Stage 2: Locking, passive movement with triggering;, or Stage 3: Locked. A satisfactory result was noted in 96 % of cases at follow-up (mean 44 months), while complete recovery was noted in 27 % of thumbs at follow-up (mean 62 months). A cure rate of 80 % was reported for Stage 2 thumbs at follow-up (mean 56 months) and 25 % for Stage 3 thumbs at follow-up (mean 68 months). The cure rate for initial Stage 2 thumbs was significantly higher than for initial Stage 3 thumbs ($p < 0.05$) [116].

In a similar prospective, consecutive case series, Jung et al. [117] examined treatment with PROM only in children ($n=30$), thumbs ($n=35$). PROM was applied 10–20 times per day. Digits were categorized as: Grade OA, extension beyond 0° without triggering; Grade OB, extension to 0° without triggering; Grade 1 active extension with triggering; Grade 2 passive extension with triggering; and Grade 3, locked. Pretest results found thumbs were identified as: Grade 1, 6 thumbs (17 %); Grade 2, 25 thumbs (71 %); and Grade 3, 3 thumbs (25 %). Posttest results found thumbs were identified as: Grade OA, 7 thumbs (20 %); Grade OB, 25 thumbs (21 %); Grade I, 5 thumbs (14 %); Grade II, 2 thumbs (6 %); No change = 1 thumb. The researchers found children with bilateral trigger thumb and children with a Grade III thumb were more likely to have an unfavorable outcome. Passive ROM seems useful for Grades 1 and 2, but may not be useful for Grade 3 trigger thumb. Additionally, PROM may be useful to correct deformity but triggering may persist [117]. These studies provide limited to moderate support for use of PROM to reduce triggering and improve motion for children with trigger thumb.

Orthotics

Two studies have described the effectiveness of orthoses to treat trigger thumb with varying outcomes [118, 119]. Koh et al. [118] conducted a retrospective, non-randomized, controlled study by reviewing medical records of children with locked interphalangeal (IP) joint. Parents self-selected whether to have their child wear an orthosis ($n=26$) or undergo observation alone ($n=38$). Children receiving a custom made, coil orthosis to hold the IP joint in extension while preventing hyperextension of the MCP joint wore the orthosis at night. Duration of treatment or observation was individualized until either resolution was achieved or surgery was indicated. The targeted outcome was full AROM of the thumb IP joint without snapping but no measurement technique was described. Of patients treated with an orthosis, 92 % experienced complete resolution within 22 months, whereas 60 % in the observation group had complete resolution in 59 months. After an additional 11 months, 97 % of patients in the observation group experienced resolution of snapping. All patients in both groups experienced complete resolution, but four (two from each group) required surgery due to continued snapping. Those receiving an orthosis had significantly higher rates of resolution ($p<0.05$) and shorter resolution time ($p<0.01$) compared to observation alone. This study suggests patients with locked trigger thumbs who wear a coil orthosis may have faster rates of resolution compared to those receiving no treatment [118]. Using a similar design, Lee et al. [119] compared treatment with an orthosis to observation alone for management of trigger thumb. In this non-randomized, non-blinded, and case controlled study, parents of children self-selected to receive an orthosis ($n=31$ thumbs) or be observed ($n=31$ thumbs). An orthosis that maintained the MCP joint and IP joint in extension was custom fabricated from thermoplastic for patients in the orthosis group, and was to be worn all day for 6–12 weeks in the child's usual environments. The orthosis was worn at night only once active extension was achieved. Mean duration of treatment was 11.7 weeks ± 6.6 weeks. The outcome classification was cured (full AROM), improved (full AROM with snapping less than once per week), or non-improved (persistent flexion deformity or surgery was requested). Regarding AROM, no measurement technique was described. In the group that received an orthosis, 12 were cured, 10 were improved, and nine were non-improved. In the observation alone group four were cured, three were improved, and 24 were unimproved. The difference between the groups was statistically significant ($p<0.05$). Response rates were 71 % for the orthosis group and 23 % for the observation alone group [119]. These studies provide limited support for use of an orthosis to manage trigger thumb when conservative treatment is desired.

References

1. Lamb DW. Radial club hand. J Bone Joint Surg. 1977;59A:1–13.
2. Brooks C. Rehabilitation of radial club hand. Tech Hand Up Extrem Surg. 1998;2(1):78–85.
3. Lake A. Hand therapy for children with congenital hand differences. Tech Hand Up Extrem Surg. 2010;14(2):78–84.
4. Holtslag I, van Wijk I, Hartog H, van der Molen AM, van der Sluis C. Long-term functional outcome of patients with longitudinal radial deficiency: cross-sectional evaluation of function, activity and participation. Disabil Rehabil. 2013;35(16):1401–7.
5. de Jong IG, Reinders-Messelink HA, Tates K, Janssen WG, Poelma MJ, van Wijk I, van der Sluis CK. Activity and participation of children and adolescents with unilateral congenital below elbow deficiency: an online focus group study. J Rehabil Med. 2012;44(10):885–92.
6. Kotwal PP, Varshney MK, Soral A. Comparison of surgical treatment and nonoperative management for radial longitudinal deficiency. J Hand Surg Eur Vol. 2012;37(2):161–9.
7. Buffart LM, Roebroeck ME, Janssen WG, Hoekstra A, Selles RW, Hovius SE, Stam HJ. Hand function and activity performance of children with longitudinal radial deficiency. J Bone Joint Surg Am. 2008;90(11):2408–15.
8. Ekblom AG, Dahlin LB, Rosberg HE, Wiig M, Werner M, Arner M. Hand function in children with radial longitudinal deficiency. BMC Musculoskelet Disord. 2013;14:116.
9. Soucie JM, Wang C, Forsyth A, Funk S, Denny M, Roach KE, Boone D. Range of motion measurements: reference values and a database for comparison. Haemophilia. 2011;17(3):500–7.
10. Barad JH, Kim RS, Ebramzadeh E, Silva M. Range of motion of the healthy pediatric elbow: cross-sectional study of a large population. J Pediatr Orthop B. 2013;22(2):117–22.
11. de Kraker MD, Selles RW, Molenaar TM, Schreuders TA, Hovius SE, Stam HJ. Palmar abduction measurements: reliability and introduction of normative data in healthy children. J Hand Surg Am. 2009;34A:1704–8.
12. Häger-Ross C, Rösblad B. Norms for grip strength in children aged 4–16. Acta Paediatr. 2002;91(6):617–25.

13. Bowman OJ, Katz B. Hand strength and prone extension in right-dominant, 6 to 9 year olds. Am J Occup Ther. 1984;38(6):367–76.
14. Fullwood D. Australian norms for hand and finger strength of boys and girls aged 5–12 years. Aust Occup Ther J. 1986;33:26–37.
15. De Smet L, Vercamment A. Grip strength in children. J Pediatr Orthop. 2001;10(4):352–4.
16. Holm I, Fredriksen P, Fosdahl M, Vøllestad N. A normative sample of isotonic and isokinetic muscle strength measurements in children 7 to 12 years of age. Acta Paediatr. 2008;97(5):602–7.
17. Ploegmakers JJ, Hepping AM, Geertzen JH, Bulstra SK, Stevens M. Grip strength is strongly associated with height, weight and gender in childhood: a cross sectional study of 2241 children and adolescents providing reference values. J Physiother. 2013;59(4):255–61.
18. Mathiowetz V, Wiemer DM, Federman SM. Grip and pinch strength: norms for 6- to 19-year-olds. Am J Occup Ther. 1986; 40(10):705–11.
19. Lee-Valkov PM, Aaron DH, Eladoumikdachi F, Thornby J, Netscher DT. Measuring normal hand dexterity values in normal 3-, 4-, and 5-year-old children and their relationship with grip and pinch strength. J Hand Ther. 2003;16(1):22–8.
20. Ager CL, Olivett BL, Johnson CL. Grasp and pinch strength in children 5 to 12 years old. Am J Occup Ther. 1984;38(2): 107–13.
21. Surrey LR, Hodson J, Robinson E, Schmidt S, Schulhof J, Stoll L, Wilson-Diekhoff N. Pinch strength norms for 5-to 12-year-olds. Phys Occup Ther Pediatr. 2001;21(1):37–49.
22. De Smet L, Decramer A. Key pinch force in children. J Pediatr Orthop B. 2006;15(6):426–7.
23. Cope EB, Antony JH. Normal values for the two-point discrimination test. Pediatr Neurol. 1992;8(4):251–4.
24. Mathiowetz V, Federman SM, Wiemer DM. Box and blocks test of manual dexterity: norms for 6–19 year olds. Can J Occup Ther. 1985;52(5):241–5.
25. Gogola GR, Velleman PF, Xu S, Morse AM, Lacy B, Aaron D. Hand dexterity in children: administration and normative values of the functional dexterity test. J Hand Surg Am. 2013;38(12):2426–31.
26. Smith YA, Hong E, Presson C. Normative and validation studies of the Nine-hole Peg Test with children. Percept Mot Skills. 2000;90(3 Pt 1):823–43.
27. Poole JL, Burtner PA, Torres TA, McMullen CK, Markham A, Marcum ML, et al. Measuring dexterity in children using the Nine-hole Peg Test. J Hand Ther. 2005;18(3):348–51.
28. Wilson BC, Iacoviello JM, Wilson JJ, Risucci D. Purdue Pegboard performance of normal preschool children. J Clin Neuropsychol. 1982;4(1):19–26.
29. Skerik SK, Weiss MW, Flatt AE. Functional evaluation of congenital hand anomalies. Am J Occup Ther. 1971;25(2):98–104.
30. Percival NJ, Sykes PJ, Chandraprakasam T. A method of assessment of pollicisation. J Hand Surg Br. 1991;16(2):141–3.
31. Ho ES, Clarke HM. Upper extremity function in children with congenital hand anomalies. J Hand Ther. 2005;18(3):352–64.
32. Buffart LM, Roebroeck ME, Pesch-Batenburg JM, Janssen WG, Stam HJ. Assessment of arm/hand functioning in children with a congenital transverse or longitudinal reduction deficiency of the upper limb. Disabil Rehabil. 2006;28(2):85–95.
33. Buffart LM, Roebroeck ME, Janssen WG, Hoekstra A, Hovius SE, Stam HJ. Comparison of instruments to assess hand function in children with radius deficiencies. J Hand Surg Am. 2007;32(4):531–40.
34. Kaplan JD, Jones NF. Outcome measures of microsurgical toe transfers for reconstruction of congenital and traumatic hand anomalies. J Pediatr Orthop. 2014;34:362–8.
35. Arnould C, Penta M, Renders A, Thonnard JL. ABILHAND-Kids: a measure of manual ability in children with cerebral palsy. Neurology. 2004;63(6):1045–52.
36. Krumlinde-Sundholm L, Holmefur M, Kottorp A, Eliasson AC. The Assisting Hand Assessment: current evidence of validity, reliability, and responsiveness to change. Dev Med Child Neurol. 2007;49(4):259–64.
37. Holmefur M, Krumlinde-Sundholm L, Eliasson AC. Interrater and intrarater reliability of the Assisting Hand Assessment. Am J Occup Ther. 2007;61(1):79–84.
38. Holmefur M, Aarts P, Hoare B, Krumlinde-Sundholm L. Test-retest and alternate forms reliability of the assisting hand assessment. J Rehabil Med. 2009;41(11):886–91.
39. Skold A, Hermansson LN, Krumlinde-Sundholm L, Eliasson AC. Development and evidence of validity for the Children's Hand-use Experience Questionnaire (CHEQ). Dev Med Child Neurol. 2011;53(5):436–42.
40. Varni JW, Seid M, Kurtin PS. PedsQL 4.0: reliability and validity of the Pediatric Quality of Life Inventory version 4.0 generic core scales in healthy and patient populations. Med Care. 2001;39(8): 800–12.
41. Schlenz AM, Schatz J, McClellan CB, Roberts CW. Responsiveness of the PedsQL to pain-related changes in health-related quality of life in pediatric sickle cell disease. J Pediatr Psychol. 2012;37(7):798–807.
42. Haley SM, Coster WJ, Faas RM. A content validity study of the Pediatric Evaluation of Disability Inventory. Pediatr Phys Ther. 1991;3:177–84.
43. Feldman AB, Haley SM, Coryell J. Concurrent and construct validity of the Pediatric Evaluation of Disability Inventory. Phys Ther. 1990;70(10):602–10.
44. Wright FV, Boschen KA. The Pediatric Evaluation of Disability Inventory (PEDI): validation of a new functional assessment outcome instrument. Can J Rehabil. 1993;7:41–2.
45. Nichols DS, Case-Smith J. Reliability and validity of the Pediatric Evaluation of Disability Inventory. Pediatr Phys Ther. 1996;8: 15–24.
46. Daltroy LH, Liang MH, Fossel AH, Goldberg MJ. The POSNA pediatric musculoskeletal functional health questionnaire: report on reliability, validity and sensitivity to change. J Pediatr Orthop. 1998;18(5):561–71.
47. Pencharz J, Young NL, Owen JL, Wright JG. Comparison of three outcomes instruments in children. J Pediatr Orthop. 2001;21(4): 425–32.
48. Amor CJ, Spaeth MC, Chafey DH, Gogola GR. Use of the Pediatric Outcomes Data Collection Instrument to evaluate functional outcomes in arthrogryposis. J Pediatr Orthop. 2011;31(3):293–6.
49. Haynes R, Sullivan E. The Paediatric Orthopaedic Society of North America paediatric orthopaedic functional health questionnaire: an analysis of normal. J Pediatr Orthop. 2001;21(5):619–21.
50. Wright FV, Hubbard S, Naumann S, Jutai J. Evaluation of the validity of the prosthetic upper extremity functional index for children. Arch Phys Med Rehabil. 2003;84(4):518–27.
51. Wright FV, Hubbard S, Jutai J, Naumann S. The Prosthetic Upper Extremity Functional Index: development and reliability testing of a new functional status questionnaire for children who use upper extremity prostheses. J Hand Ther. 2001;14(2):91–104.
52. Bagley AM, Molitor F, Wagner LV, Tomhave W, James MA. The unilateral below elbow test: a function test for children with unilateral congenital below elbow deficiency. Dev Med Child Neurol. 2006;48(7):569–75.
53. Netscher DT, Aliu O, Sandvall BK, Staines KG, Hamilton KL, Salazar-Reyes H, Thornby J. Functional outcomes of children with index pollicizations for thumb deficiency. J Hand Surg Am. 2013;38(2):250–7.
54. Ardon MS, Selles RW, Roebroeck ME, Hovius SE, Stam HJ, Janssen WG. Poor agreement on health-related quality of life between children with congenital hand differences and their parents. Arch Phys Med Rehabil. 2012;93(4):641–6.

55. Sheffler LC, Hanley C, Bagley A, Molitor F, James MA. Comparison of self-reports and parent proxy-reports of function and quality of life of children with below-the-elbow deficiency. J Bone Joint Surg Am. 2009;91(12):2852–9.
56. Giele H, Giele C, Bower C, Allison M. The incidence and epidemiology of congenital upper limb anomalies: a total population study. J Hand Surg Am. 2001;26(4):628–34.
57. Koskimies E, Lindfors N, Gissler M, Peltonen J, Nietosvaara Y. Congenital upper limb deficiencies and associated malformations in Finland: a population-based study. J Hand Surg Am. 2011;36(6):1058–65.
58. Ekblom AG, Laurell T, Arner M. Epidemiology of congenital upper limb anomalies in 562 children born in 1997 to 2007: a total population study from Stockholm, Sweden. J Hand Surg Am. 2010;35(11):1742–54.
59. Froster-Iskenius UG, Baird PA. Limb reduction defects in over one million consecutive live births. Teratology. 1998;39(2):127–35.
60. Evans JA, Vitez M, Czeizel A. Congenital abnormalities associated with limb deficiency defects: a population study based on cases from the Hungarian Congenital Malformation Registry (1975-1984). Am J Med Genet. 1994;49(1):52–66.
61. Baker RP, Field J, Gozzard C, Wyatt MC, Robertson Y. Does postoperative hand elevation reduce swelling? A randomized study. J Hand Surg Eur Vol. 2010;35(3):192–4.
62. Fagan DJ, Evans A, Ghandour A, Prabhkaran P, Clay NR. A controlled clinical trial of postoperative hand elevation at home following day case surgery. J Hand Surg Br. 2004;29:458–60.
63. Enos L, Lane R, MacDougal BA. Brief or new: the use of self-adherent wrap in hand rehabilitation. Am J Occup Ther. 1984;38(4):265–6.
64. Lowell M, Pirc P, Ward RS, et al. Effect of 3 M Coban self-adherent wraps on edema and function of the burned hand: a case study. J Burn Care Rehabil. 2003;24:253–8.
65. Shin TM, Bordeaux JS. The role of massage in scar management: a literature review. Dermatol Surg. 2012;38(3):414–23.
66. Foo CW, Tristani-Firouzi P. Topical modalities for treatment and prevention of postsurgical hypertrophic scars. Facial Plast Surg Clin North Am. 2011;19(3):551–7.
67. Chang LW, Deng WP, Yeong EK, Wu CY, Yeh SW. Pressure effects on the growth of human scar fibroblasts. J Burn Care Res. 2008;29(5):835–41.
68. Kischer CW, Shetlar MR, Shetlar CL. Alteration of hypertrophic scars induced by mechanical pressure. Arch Dermatol. 1975;111(1):60–4.
69. Anzarut A, Olson J, Singh P, Rowe BH, Tredget EE. The effectiveness of pressure garment therapy for the prevention of abnormal scarring after burn injury: a meta-analysis. J Plast Reconstr Aesthet Surg. 2009;62(1):77–84.
70. Steinstraesser L, Flak E, Witte B, Ring A, Tilkorn D, Hauser J, Langer S, Steinau HU, Al-Benna S. Pressure garment therapy alone and in combination with silicone for the prevention of hypertrophic scarring: randomized controlled trial with intraindividual comparison. Plast Reconstr Surg. 2011;128(4):306e–13.
71. Widgerow AD. Cellular/extracellular cross-talk in scar evolution and control. Wound Repair Regen. 2011;19:117–33.
72. Hart RG, Wolff TW, O'Neil WL. Preventing tourniquet effect when dressing finger wounds in children. Am J Emerg Med. 2004;22:594–5.
73. Yelvington M, Brown S, Castro MM, Nick TG. The use of neoprene as a scar management modality. Burns. 2013; pii:S0305-4179(12)00364-6.
74. Mustoe TA, Cooter RD, Gold MH, Hobbs FD, Ramelet AA, Shakespeare PG, Stella M, Téot L, Wood FM, Ziegler UE, International Advisory Panel on Scar Management. International clinical recommendations on scar management. Plast Reconstr Surg. 2002;110(2):560–71.
75. O'Brien L, Pandit A. Silicon gel sheeting for preventing and treating hypertrophic and keloid scars. Cochrane Database Syst Rev. 2006;1, CD003826.
76. O'Brien L, Jones DJ. Silicone gel sheeting for preventing and treating hypertrophic and keloid scars. Cochrane Database Syst Rev. 2013;9, CD003826.
77. Sawada Y, Sone K. Hydration and occlusion treatment for hypertrophic scars and keloids. Br J Plast Surg. 1992;45(8):599–603.
78. de Oliveira GV, Nunes TA, Magna LA, Cintra ML, Kitten GT, Zarpellon S, Raposo Do Amaral CM. Silicone versus nonsilicone gel dressings: a controlled trial. Dermatol Surg. 2001;27(8):721–6.
79. Rustad KC, Wong VW, Gurtner GC. The role of focal adhesion complexes in fibroblast mechanotransduction during scar formation. Differentiation. 2013;86(3):87–91.
80. Chiquet M, Gelman L, Lutz R, Maier S. From mechanotransduction to extracellular matrix gene expression in fibroblasts. Biochim Biophys Acta. 2009;1793:911–20.
81. Widgerow AD, Chait LA, Stals PJ, Stals R, Candy G. Multimodality scar management program. Aesthetic Plast Surg. 2009;33:533–43.
82. Atkinson JM, McKenna KT, Barnett AG, McGrath DJ, Rudd M. A randomized, controlled trial to determine the efficacy of paper tape in preventing hypertrophic scar formation in surgical incisions that traverse Langer's skin tension lines. Plast Reconstr Surg. 2005;116(6):1648–56.
83. Widgerow AD, Chait LA. Scar management practice and science: a comprehensive approach to controlling scar tissue and avoiding hypertrophic scarring. Adv Skin Wound Care. 2011;24(12):555–61.
84. Michlovitz SL, Harris BA, Watkins MP. Therapy interventions for improving joint range of motion: a systematic review. J Hand Ther. 2004;17(2):118–31.
85. Glasgow C, Tooth LR, Fleming J. Mobilizing the stiff hand: combining theory and evidence to improve clinical outcomes. J Hand Ther. 2010;23(4):392–400.
86. Flowers KR. A proposed decision hierarchy for splinting the stiff joint, with an emphasis on force application parameters. J Hand Ther. 2002;15(2):158–62.
87. Weeks PM, Wray RC. Management of acute hand injuries. 2nd ed. St. Louis: Mosby; 1978. p. 434–8.
88. Flowers KR, LaStayo PC. Effect of total end range time on improving passive range of motion. J Hand Ther. 1994;7(3): 150–7.
89. Glasgow C, Wilton J, Tooth L. Optimal daily total end range time for contracture: resolution in hand splinting. J Hand Ther. 2003;16(3):207–18.
90. Glasgow C, Fleming J, Tooth LR, Peters S. Randomized controlled trial of daily total end range time (TERT) for capener splinting of the stiff proximal interphalangeal joint. Am J Occup Ther. 2012;66(2):243–8.
91. Blair WF, Shurr DG, Buckwalter JA. Functional status in ulnar deficiency. J Pediatr Orthop. 1983;3(1):37–40.
92. Ty J, James MA. Failure of differentiation: Part II (arthrogryposis, camptodactyly, clinodactyly, Madelung deformity, trigger finger, and trigger thumb). Hand Clin. 2009;25(2):195–214.
93. Rhee SH, Oh WS, Lee HJ, Roh YH, Lee JO, Baek GH. Effect of passive stretching on simple camptodactyly in children younger than three years of age. J Hand Surg Am. 2010;35(11):1768–73.
94. Hori M, Nakamura R, Inoue G, Imamura T, Horii E, Tanaka Y, Miura T. Nonoperative treatment of camptodactyly. J Hand Surg Am. 1987;12(6):1061–5.
95. Miura T, Nakamura R, Tamura Y. Long-standing extended dynamic splintage and release of an abnormal restraining structure in camptodactyly. J Hand Surg Br. 1992;17(6):665–72.
96. Benson LS, Waters PM, Kamil NI, Simmons BP, Upton 3rd J. Camptodactyly: classification and results of nonoperative treatment. J Pediatr Orthop. 1994;14(6):814–9.

97. de Roode CP, James MA, McCarroll HR. Abductor digit minimi opponensplasty: technique, modifications, and measurement of opposition. Tech Hand Up Extrem Surg. 2010;14(1):51–3.
98. Goldfarb CA, Calhoun VD, Daily L, Manske PR. Hand and upper extremity therapy: congenital, pediatric and adolescent: St Louis protocols. St. Louis: Shriners Hospital for Children; 2011.
99. Egerszegi EP. Reconstruction of congenital hand anomalies to provide stable pinch and/or grasp: case pictorial series. J Pediatr Rehabil Med. 2009;2(3):173–9.
100. Kozin SH, Ezaki M. Flexor digitorum superficialis opponensplasty with ulnar collateral ligament reconstruction for thumb deficiency. Tech Hand Up Extrem Surg. 2010;14(1):46–50.
101. Manske PR, Goldfarb CA. Congenital failure of formation of the upper limb. Hand Clin. 2009;25(2):157–70.
102. Bednar MS, James MA, Light TR. Congenital longitudinal deficiency. J Hand Surg Am. 2009;34(9):1739–47.
103. Damore E, Kozin SH, Thoder JJ, Porter S. The recurrence of deformity after surgical centralization for radial clubhand. J Hand Surg Am. 2000;25(4):745–51.
104. Banskota AK, Bijukachhe B, Rajbhandary T, Pradhan I, Singh A. Radial club hand deformity—the continuing challenges and controversies. Kathmandu Univ Med J (KUMJ). 2005;3(1):30–4.
105. Fuller M. Treatment of congenital differences of the upper extremity: therapists commentary. J Hand Ther. 1999;12(2):174–7.
106. Van Heest A. Wrist centralization using the dorsal rotation flap in radial longitudinal deficiency. Tech Hand Up Extrem Surg. 2010;14(2):94–9.
107. Wall LB, Ezaki M, Oishi SN. Management of congenital radial longitudinal deficiency: controversies and current concepts. Plast Reconstr Surg. 2013;132(1):122–8.
108. Kennedy SM. Neoprene wrist brace for correction of radial club hand in children. J Hand Ther. 1996;9(4):387–90.
109. Deunk J, Nicolai JP, Hamburg SM. Long-term results of syndactyly correction: full-thickness versus split-thickness skin grafts. J Hand Surg Br. 2003;28(2):125–30.
110. Lumenta DB, Kitzinger HB, Beck H, Frey M. Long-term outcomes of web creep, scar quality, and function after simple syndactyly surgical treatment. J Hand Surg Am. 2010;35(8):1323–9.
111. Vekris MD, Lykissas MG, Soucacos PN, Korompilias AV, Beris AE. Congenital syndactyly: outcome of surgical treatment in 131 webs. Tech Hand Up Extrem Surg. 2010;14(1):2–7.
112. Mallet C, Ilharreborde B, Jehanno P, Litzelmann E, Valenti P, Mazda K, Penneçot GF, Fitoussi F. Comparative study of 2 commissural dorsal flap techniques for the treatment of congenital syndactyly. J Pediatr Orthop. 2013;33(2):197–204.
113. Goldfarb CA, Steffen JA, Stutz CM. Complex syndactyly: aesthetic and objective outcomes. J Hand Surg Am. 2012;37(10):2068–73.
114. Moran SL, Tomhave W. Management of congenital hand anomalies. In: Skirven TM, Osterman AL, Fedorzyk JM, Amadio PC, editors. Rehabilitation of the hand and upper extremity. 6th ed. Philadelphia: Elsevier Mosby; 2011. p. 1631–50.
115. Baek GH, Lee HJ. The natural history of pediatric trigger thumb: a study with a minimum of five years follow-up. Clin Orthop Surg. 2011;3(2):157–9.
116. Wantabe H, Hamada Y, Toshima T, Nagasawa K. Conservative treatment for trigger thumb in children. Arch Orthop Trauma Surg. 2001;121(7):388–90.
117. Jung HJ, Lee JS, Song KS, Yang JJ. Conservative treatment of pediatric trigger thumb: follow-up for over 4 years. J Hand Surg Eur Vol. 2011;37E(3):220–4.
118. Koh S, Horii E, Hattori T, Hiroishi M, Otsula J. Pediatric trigger thumb with locked interphalangeal joint: can observation or splinting be a treatment option? J Pediatr Orthop. 2012;32(7):724–6.
119. Lee Z, Chang C, Yang W, Hung S, Shih C. Extension splint for trigger thumb in children. J Pediatr Orthop. 2006;26(6):785–7.