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Congenital thumb anomalies and the consequences for daily life: patients’
long-term experience after corrective surgery. A qualitative study.

Running Title: Congenital thumb anomalies – consequences for daily life.

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Keywords: Thumb hypoplasia/aplasia, thumb duplication, pollicisation, hand
disfigurement, adaptation, qualitative research

24 **ABSTRACT**

25 *Purpose:* The aim of the study was to explore patients' long-term experience of a
26 congenital hand problem, and the consequences for daily life.

27 *Method:* Fifteen participants with a median age 24 years (17-55), born with thumb
28 hypoplasia/aplasia or thumb duplication were interviewed using a semi-structured
29 interview guide. The interviews were subjected to qualitative content analysis.

30 *Results:* Although the mobility and strength in the thumb/hand(s) varied within the
31 group, hand function was generally described as good. Compensatory strategies were
32 used to overcome practical obstacles. The emotional reactions to being visibly different
33 from peers in early life varied from total acceptance and a sense of pride in being
34 special, to deep distress and social withdrawal. Support from parents, teachers and
35 others was important in facing emotional challenges and practical consequences.

36 *Conclusion:* The present study highlights the importance of healthcare professionals
37 addressing appearance-related concerns which may have long-term emotional and social
38 consequences for patients born with a thumb anomaly.

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46 **Implications for Rehabilitation**

47 · Appearance related concerns and need for emotional support should be fully

48 considered throughout the rehabilitation process to prevent distress and social

49 withdrawal

50 · Effective problem-solving strategies, such as compensation, change in occupational

51 performance and support from others may reduce activity limitations and participation

52 restriction.

53

54 INTRODUCTION

55 Congenital malformations in the upper extremities, with an overall reported incidence of
56 21.5/10 000 live births [1], may have serious consequences for the individual patient.

57 A variety of such malformations involve the thumb, causing possible functional
58 disturbances of varying severity as the thumb is crucial for proper hand function.

59 Thumb hypoplasia/aplasia, with a “non-functioning”, thumb represents a surgical
60 challenge and usually requires a pollicisation procedure, while thumb duplication poses
61 minor surgical and functional problems in the hand.

62 Earlier reports have mainly focused on quantitative research, such as technical issues,
63 functional outcome and subjective assessment of appearance, in both thumb hypoplasia
64 and thumb duplication [2-5]. However, long-term satisfaction and well-being in patients
65 with e.g. thumb hypoplasia/aplasia and thumb duplication may depend on several other
66 factors such as emotional stress related to functional limitations or the experience of
67 being visibly different [6]. Such emotional stress throughout childhood has been
68 reported to affect a majority of children with congenital hand differences and may not
69 always be recognized by parents or caregivers [6]. Access to effective coping
70 mechanisms to counteract stress factors when facing challenges in daily life may also
71 vary. It is therefore important to further illuminate the long-term consequences in daily
72 life for this patient group.

73 A qualitative approach with open questions may deepen the understanding of how
74 patients perceive and handle such consequences in daily life [6,7]. Our aim was to
75 explore the patient’s experience of a congenital hand problem, focusing on thumb
76 hypoplasia/aplasia and thumb duplication, and the long-term consequences for daily
77 life, including emotional and social aspects as well as impact on activity and

78 participation. We were also interested in personal qualities and the support needed to
79 achieve positive adaptation.

80

81

82 **METHOD**

83 **Design and Participants**

84 A qualitative descriptive method with an inductive approach was used to reach a deeper
85 understanding about the consequences for the individual of being born with a congenital
86 hand problem. In total, 15 individuals were included and interviewed, seven with a
87 thumb hypoplasia and eight with a duplicated thumb. The sample size was determined
88 by the role of redundancy, meaning that the last four interviews provided little new
89 information [7].

90 All participants were patients admitted to the Hand Surgery Clinic, Skane University
91 Hospital, between the years 1978-1998 who could be interviewed (i.e. had no serious
92 mental, cognitive, and or linguistic impairments). Twenty-two patients were eligible for
93 inclusion. Seven patients were not included as they declined participation due to lack of
94 time or living too far away or could not be reached. The thumb duplications included
95 were of the Wassel IV [8] and the thumb aplasia was Type IIIB, IV or V [9,10] and all
96 had had previous surgery. Five of the patients with thumb hypoplasia had undergone a
97 pollicisation procedure, two had tendon transfers and one had only removal of the
98 rudimentary thumbs [11]. The patients with thumb duplication had surgery as described
99 elsewhere for this type [12].

100 The median age was 24 years (17-55). All patients except one had finished high school,
101 four were in secondary education and three were completing their university studies.

102 One patient was unemployed and seven patients worked in a variety of professions,
103 such as sales, economy, project management, professional driving, restaurant work and
104 healthcare professions. Six participants lived with their parents, five had a partner and
105 four had a family with children. Seven men and eight women participated. Table 1.
106 Prior to the interview the participants completed the Swedish versions of several self-
107 report questionnaires; the QuickDASH [13], the condensed 13-item Sense of Coherence
108 (SOC) [14], the acute version of the Short Form 36 (SF-36) questionnaire [15], the Cold
109 Intolerance Symptom Severity (CISS) questionnaire [16,17] and seven single questions
110 (numeric rating scale 0-100, 0 representing no problem and 100 worst possible problem)
111 concerning pain at rest, pain during motion without load, pain with load, grip function,
112 fine motor skill, weakness and appearance of the hand. The interviewer also measured
113 grip strength, key pinch and pinch strength [18,19]. Table 2.

114

115 **Procedure and ethics**

116 Written information was sent to the participants emphasizing the voluntary nature of the
117 study. The first author then contacted them and arranged an interview time for those
118 who agreed to participate. Written consent was obtained in conjunction with the
119 interview and all participants were informed about how the data would be analysed and
120 were assured of confidentiality. The collection of all data was conducted by the first
121 author not previously involved with the participants care. The study was performed in
122 accordance with the ethical guidelines stated in the Helsinki Declaration and the
123 Swedish Act Concerning the Ethical Review of Research Involving Humans
124 (SFS:2003:460) and approved by the local ethics committee of Lund University (Dnr
125 2009/339). All interviews were performed and tape-recorded by the first author in a

126 quiet room at the clinic and lasted between 20 and 66 minutes. The interview started
127 with a repetition of the aim of the study. A semi-structured interview guide with open
128 questions was then used and the participants were asked to describe their overall
129 thumb/hand function, pain, appearance, emotional and social consequences, personal
130 qualities, support from others and the impact on activity and participation. Follow-up
131 questions were asked such as: How did you experience that? How did you handle that?
132 Can you describe that in more detail? A secretary, marking nonverbal expressions, then
133 transcribed all the interviews verbatim. All transcripts were checked for accuracy by the
134 first author, who also translated the quotations from Swedish into English. The
135 translations were verified by the last author.

136

137 **Data analysis**

138 The text was read and reread by the first and last author and subjected to qualitative
139 content analysis [20,7]. The analysis started with a naive reading of each interview to
140 gain a general impression of the content. Meaning units, described as words or
141 sentences related to each other through their content and related to the aim of the study,
142 were then identified. The impression of the text was discussed and the selected meaning
143 units compared. The meaning units were shortened into codes (labels for the meaning
144 units) and then grouped into categories. Within each category similar statements were
145 analysed critically and questioned, then read and compared until a reasonable
146 interpretation was reached. The categories were then discussed with the second author
147 and adjustments were made to ensure that the categories covered all aspects in the text.
148 Finally, the categories were compared with the text and with each other. The second
149 author read seven randomly selected interviews and reviewed the different codes and

150 categories. Concerning the authors' pre-understanding, the first author is an experienced
151 occupational therapist specialized in hand rehabilitation; the second and last authors are
152 experienced hand surgeons. All three authors work in a specialized unit. Both the first
153 and the second authors are experienced in qualitative research methodology [21-24].

154

155

156 **RESULTS**

157 **Consequences for hand function.**

158 Even though the mobility in the affected thumb(s) varied among participants they
159 described normal sensibility and overall good hand function. *"I have not been limited –*
160 *there is just one finger less..." (Participant 12)*. However, situations requiring fine
161 motor skills and dexterity could be troublesome, e.g. when undoing a necklace or
162 putting on earrings. Participants who had undergone a pollicisation procedure also
163 described a sense of weakness and tiredness requiring the use of modified grip patterns
164 using other fingers or the unaffected side or both hands. Depending on the muscle
165 strength, those born with duplicated thumbs used similar habitual strategies. *"It works*
166 *well; I was born with the problem so I have learned to compensate by using both hands*
167 *when needed"*. (Participant 10).

168 With age, hand function and dealing with everyday challenges were considered
169 increasingly important compared to a visibly different appearance.

170

171 **Pain experience**

172 Pain at rest was uncommon, but hitting the base of the thumb or carrying something
173 heavy could trigger pain. This was described as an increasing pain, a sense of strain

174 rather than actual pain or as if the lack of muscles enabled the pain to continue deep
175 inside, triggering numbness and an ache in the bones. *“The whole hand explodes and*
176 *then it feels a little numb...”* (Participant 1). The pain could be unbearable, like having
177 a knife or a nail in the hand and, if severe, affecting sleep. A premonition of pain was
178 also connected to locking within the wrist, or present in the unaffected hand when it was
179 overused and when the other hand worked mainly as a support hand. Compensatory grip
180 patterns and use of the whole body, technical aids or simply enduring the pain were
181 strategies used. *“I squeeze my hand but the hand doesn’t listen. I try to massage the*
182 *hand to alleviate the pain.”* (Participant 9). For those with bilateral anomalies and
183 constant pain in both hands the possibility of using an unaffected side to compensate
184 was lost, which caused distress and worries for the future. Re-education and
185 occupational changes was therefore necessary. *“In the morning I go straight to the*
186 *medicine (paracetamol) and in the evening and four times daily. I can’t cope. It is*
187 *frightening.”* (Participant 14). Exposure to cold caused pain, stiffness, numbness and a
188 change in skin colour for those affected and was experienced as a numb feeling deep
189 inside, as if the thumb was lost and had been cut off.

190

191 **Consequences for activity and participation**

192 When grip function, fine motor skills and weakness were affected the participants
193 described fruitful strategies for overcoming challenges, and engagement in activities
194 and participation in society were realizable. In school, writing could cause pain in the
195 thumb and required the use of specially adapted pencils or breaks. This was especially
196 apparent under time pressure and in examinations, both at younger ages and during
197 university education. Various activities in physical education (gymnastics), such as

198 hand-, basket-, or volley-ball, push-ups, cartwheels, turning upside down/handstands,
199 grasping a bar, could trigger pain or cramps or were impossible for some because of
200 reduced strength. In textile handicraft the ability to handle a needle and thread required
201 dexterity skills and small needles were sometimes difficult to grip because of reduced
202 strength. Supportive and understanding teachers were important in finding solutions or
203 alternative activities.

204 Activities at home, such as holding on to a broom or vacuum cleaner, opening a jar,
205 peeling potatoes, using cutlery or scissors, holding a glass or unbuttoning a pair of jeans
206 could, cause pain. A modified grip, use of the other hand or both hands, technical aids
207 or asking for help were strategies used to overcome obstacles.

208 Several leisure activities were possible to pursue and caused no problems at all. *“I play*
209 *the French horn without any problems since you don’t use the thumb while playing.”*
210 *(Participant 10). “I play the trumpet – I hold it with my left hand and play with the other*
211 *hand.” (Participant 11). “I have learned to use my right hand in a different way when I*
212 *play with the X-box, because it’s developed for five-fingered hands.” (Participant 1).*

213 However, there was also examples of the opposite, e.g. bowling put too much strain on
214 the thumb, ball sports was painful when hitting the ball, playing goal keeper in soccer
215 required too much strength and the desire to play a violin had to be abandoned in favour
216 of an accordion where a modified grip pattern enabled the person to make music.

217 *“It is not easy to go to a fitness centre to work up your strength – they don’t understand.*
218 *You are at the level of minus ten and need help to arrive at zero and the exercises are*
219 *too heavy...” (Participant 14).*

220 The career choices for those of working age were not at all or only to some extent
221 influenced by activity limitations. An altered performance, re-education or change of

222 work was alternatives followed. *“I am studying to become a chef and I can do*
223 *everything that the others do and I do it as well as them.” (Participant 1). “I had my*
224 *own tricks when turning a patient around in the bed – I rolled up the draw sheet a lot*
225 *underneath my hand and then I used my whole body and pulled.” (Participant 7).*

226

227 **Appearance - emotional consequences and social acceptance**

228 The views on the aesthetics of the thumb varied from a feeling of looking nice and
229 almost normal to ugly, scarred and misconfigured. The latter was especially
230 troublesome in childhood/adolescence, but eased during high school. *“Why don’t I look*
231 *like everybody else?” (Participant 3). Appearance-related concerns were expressed both*
232 *by those with minor or more visible disfigurements. The choice to preserve a four-*
233 *fingered hand and not proceed with a pollicisation procedure arose from the wish to*
234 *look “whole/complete”, pretending that there was a thumb hidden in the palm and*
235 *thereby looking like everyone else. It was all about hiding and, in the worst scenario,*
236 *even disappearing as a person. Habitual solutions were to hide the hand in a pocket,*
237 *wear long-sleeved sweaters or pull jacket sleeves down. Responding to questions in the*
238 *classroom by raising one’s hand was avoided because then the deformity became*
239 *visible. This could affect grades in early school years. “...sometimes I greet people and*
240 *then I hide my hand quickly so no one can tell I’m different.” (Participant 9). “Socially,*
241 *it affects me when shaking hands, because my hand is smaller and a little weird, but no*
242 *one notices, it’s all in my head.” (Participant 1). “It has become a habit that I feel*
243 *ashamed of it – everybody else has ten fingers.” (Participant 13).*

244 Intrusive or hurtful comments, such as your thumb looks like an “ape’s thumb”, what a
245 “weird thumb” you have or simply being called “CP” (person with cerebral palsy) were

246 difficult to handle as a child. The appearance of the hand gave rise to verbal teasing or
247 even bullying and caused insecurity, distress and social withdrawal.

248 *“Schoolmates in primary school commented my thumb saying - You can only count to*
249 *nine and things like that, and it makes you think – What is wrong with me?”*

250 *(Participant 12).*

251 *“ I wake up, looking at my hand, thinking – what a monster I am, remembering how I*
252 *was bullied in school and called a four-fingered freak. I had to rename myself E.T. to*
253 *save my psyche/soul... “I was very lonely, had no friends so I played with an imaginary*
254 *friend – in one respect I had two kinds of personalities...” (Participant 9). Choosing*
255 *friends carefully before opening up was a lesson learned and comparing yourself with*
256 *someone less fortunate or playing down the importance of appearance were helpful*
257 *when dealing with emotional consequences. “My father and I met another patient in*
258 *rehab who had the same problems in both hands and for me that was like, shit, I could*
259 *have had it in both hands. I was lucky, it could have been worse; I still have one hand*
260 *that works.” (Participant 1).*

261 Others experienced the opposite, feeling rather cool and special because of the thumb(s)
262 or having a hand closer to normal. Schoolmates also thought it was interesting and
263 exciting being born with e.g. duplicated thumbs. Hiding the hand was considered to
264 draw more attention to it. To “have ones place” by being physically stronger/bigger
265 instilled some fear “in” schoolmates and counteracted possible teasing or bullying.
266 Peers rarely made negative comments.

267 *“My mates thought it was a cool thing – it made me proud.” (Participant 4). “I have*
268 *never thought about it as a misconfiguration in the hand because it has always been*
269 *gone.” (Participant 6).*

270 Consequently, the emotional reactions to appearance varied from total acceptance and a
271 sense of pride in being special to deep distress and social exclusion. The bullying
272 stopped in secondary school, but earlier memories could create emotional scars resulting
273 in habitual hiding of the hand even as adults and especially among strangers. However,
274 comments referring to the smaller number of fingers on the hand could be used as
275 motivation to achieve excellent results in mathematics and with age this type of remark
276 was also considered mentally strengthening. As adults it was easier to deal with
277 whispered comments with confidence: *“Talk to me instead”*, and suddenly a new insight
278 emerged – *“I decided to put jewellery on my fingers – why should I hide myself
279 anymore.”* (Participant 14).

280

281 Approaching the other sex could be scary, especially during adolescence, even if treated
282 with respect and empathy, but with time it also became natural since being born with
283 e.g. duplicated thumbs was part of that person’s life story and was eventually accepted
284 internally. Initially, there were examples of a pattern of concealment of the disfigured
285 hand and a fear of being rejected, but the total acceptance and emotional support from
286 partners/spouses seemed to bolster confidence and self-esteem. *“When it comes to boys
287 I think a lot about, is he going to continue talking to me when he knows about the hand,
288 how will he react – it worries me a little, however, most guys seem to accept it.”*
289 (Participant 12).

290 *“A girl I met held my hand and felt that something was different but she didn’t care, she
291 just squeezed my hand tight, calmed my fear, accepted it, and then I felt that my self-
292 confidence came back to me knowing that girls could like me despite my hand.”*
293 (Participant 9).

294

295 **Support and personal characteristics**

296 The support from parents, grandparents, relatives and teachers was important when
297 facing emotional challenges and practical consequences, although support was not
298 considered a necessity for everyone. *“My father’s support has meant a great deal to me.*

299 *I told him what kind of problems I had and then we solved them together.” (Participant*
300 *9). “I have not felt the need for support – I have managed on my own.” (Participant 2).*

301 Teachers or parents gave helpful hints about compensatory measures, such as adapted
302 pencils, and how to tie shoelaces with the middle and ring finger. In school it could be
303 helpful to receive copies or assistance with writing especially after surgery when
304 strength was reduced and dexterity impaired, although it was also important for personal
305 development to manage by one’s self and not be receiving help all the time. Caring
306 parents spoke about playing down the importance of appearance, looking at the bigger
307 picture, looking outside the box and other words of wisdom.

308 *“The gym teacher knew about my hand and he was very helpful and understanding, he*
309 *took me aside and talked, not in front of everybody else because that is embarrassing*
310 *and he encouraged me to tell him if I had any problems and he would support me. I*
311 *didn’t feel any pressure or fear in gymnastics, I thought it was fun, it was my favourite*
312 *lesson.” (Participant 1).*

313 When dealing with practical and emotional challenges it was important to be self-
314 confident, positive, persistent, hard-bitten, creative, motivated and patient. To ignore
315 negative comments from schoolmates and to follow one’s own path was a fruitful
316 strategy. The strength to set limits when insulted was something that gradually
317 developed with time. Humour could be a redemptive strategy in uncomfortable

318 situations. *“I try to be funny/hilarious especially in uncomfortable situations; to joke*
319 *about it helps...” (Participant 1).* All the experiences through life were considered
320 meaningful in retrospect and were described as *“...an enormous resource that I can use*
321 *to help other people.” (Participant 9).*

322 It was important for healthcare professionals to consider the psychological aspects of
323 feeling different from others and the need to talk about it. Since the surgical procedures
324 were usually completed at an early age, our respondents stressed the importance of
325 talking directly to the child in a calm and reassuring way. Information about realistic
326 expectations concerning appearance-related improvements was also cited as important
327 as well as informative leaflets that would be useful in school.

328

329 **Advice to future patients**

330 Based on their own experiences a wealth of advice to future patients was expressed:
331 find your own way and go forward; learn to live with it, try to accept it and make the
332 best of it; don't let it hinder you; do the things that feel best for you and not what
333 pleases others; be yourself and nobody else; set your own goals; if you can't manage,
334 don't be afraid to ask for help; live a normal life and don't worry about the future.

335 *“Your hand is just like an ordinary hand, only a little weaker - you should feel special.*
336 *If someone tells you that you can't then try to prove that you actually can manage, it's*
337 *all about proving that you want it and having support from family...”(Participant 13).*

338

339

340

341

342 **DISCUSSION**

343

344 Patients' long-term experiences of being born with a congenital malformation in the
345 upper extremity (i.e. thumb hypoplasia/aplasia or thumb duplication) were highlighted
346 in this study. Although the mobility and strength in the thumb/hand(s) varied within the
347 group, hand function was generally described as good. Compensatory strategies were
348 frequently used to overcome practical obstacles caused by impaired fine motor
349 skills/dexterity, weakness or pain. The emotional reactions to being visibly different
350 from peers during early life varied from total acceptance and a sense of pride in being
351 special, to deep distress and social withdrawal. The support from parents, teachers and
352 others was important when facing emotional challenges and practical consequences.

353

354 The most striking finding in our study was the long-term psychosocial effect of being
355 visibly different, although it was not a problem for all respondents. Appearance is
356 important for an individual's self-concept and affects the way we are perceived and
357 treated by peers, teachers and others [25]. The severity or extent of a disfigurement is
358 not always related to the degree of emotional distress. A person's perception of how
359 noticeable their difference is to others is a more relevant predictor, particularly if the
360 face and hands are involved [26,27]. Hands are difficult to conceal, always on show
361 both to the child and to those with whom the child interacts. Hiding the affected hand(s)
362 in pockets or wearing long-sleeved sweaters/jackets was a strategy frequently resorted
363 to by the respondents and in the worst scenario it even brought about a wish to
364 disappear as a person. This habitual pattern was used both in school and in other social
365 contexts, especially during childhood and adolescence, but for some it extended even

366 into adulthood, despite the mildness of the visible difference. Andersson et al [28]
367 pointed out that children with milder disabilities may be at a higher risk of concealing
368 their hand and this may contribute to a poor self-concept. One explanation for this
369 might be that the hands appear closer to normal and therefore less support or sympathy
370 is received compared to children with severe visible deformities. It is impossible for
371 these children to hide the disability, they are likely to experience negative reactions
372 more consistently and the process of acceptance and effective ways of coping may
373 therefore start earlier on. This may lead to comparable self-concepts and a self-esteem
374 as high as that of healthy children [27-29]. The negative consequences, including social
375 withdrawal, for some of our participants is in line with earlier reports of lower social
376 functioning among patients with congenital hand differences, especially in older
377 children when negative self-sense and stigma may be perceived [29].

378

379 The support given by parents or close relatives was described and highly valued,
380 however, being able to talk to someone else about the psychological aspects of feeling
381 different from others was also expressed. According to Lukash, 2002, children cannot
382 adequately express their feelings about physical issues that may affect them emotionally
383 [30]. Being exposed to teasing at the vulnerable age of six or seven, when acceptance
384 from peers is of the utmost importance, may be overwhelming for the child [31]. At this
385 age children may not have access to the problem-solving skills needed for effective
386 coping and the child's reaction to negative comments may trigger and reinforce patterns
387 of bullying [31]. The opportunity to receive professional help to deal with negative
388 comments from peers and thereby bolster one's self-esteem may have facilitated
389 positive coping skills in our participants.

390 Maddern et al pointed out that a child's self-concept, including perceptions, beliefs,
391 feelings, attitudes and values about themselves, is more likely to be healthily maintained
392 if parents and teachers adopt a positive attitude towards the disfigurement [32]. It is also
393 well known that parental responses to a child's physical problem play a significant role
394 in their social well-being [31]. If comments or questions about the child's disfigurement
395 cause the parents distress, the child may feel that this is too serious a matter for their
396 parents to discuss, control and contain [31]. This was not an issue covered in depth with
397 our respondents, but an overall impression was that the support and security obtained
398 from parents, relatives and teachers was important for them to play down the
399 importance of appearance, look at the bigger picture and look outside the box. This
400 support facilitated emotion-, and problem-based coping mechanisms.

401 Some of our participants did not express any appearance-related concerns. Appearance
402 was not an issue since the hand did not differ much from normal, or even if that was the
403 case they expressed a strong self-image and described a sense of pride in being special.
404 Negative comments from peers or others rarely occurred and social interaction was
405 therefore less of a problem. Hiding the hand was considered counterproductive, drawing
406 even more attention. To minimize the importance of appearance, by e.g. comparison
407 with patients with a more visible disfigurement, had a positive affect on well-being and
408 constituted a strategy found in other studied patient groups [33]. Whether the stronger
409 resilience within these respondents is because of a personality trait enabling them to
410 access effective coping strategies, or access to social networks and other factors
411 strengthened their coping even more remains to be explored. But, as pointed out by
412 Joachim, 2000, the state of being stigmatized is more likely to depend on how the
413 individual chooses to see her/himself and not on the degree of disability [34].

414 In addition to its functional importance, the hand also has a psychological and social
415 meaning and is an instrument for sensory exploration and intimate contact [30]. During
416 adolescence appearance becomes increasingly salient. The impact of a malformation of
417 the hand, burn injury, skin disorder or cleft lip on romantic relationships and on the
418 enjoyment of an intimate relationship may be an additional concern in a challenging
419 period of life but is rarely studied [35]. However, a fear of being rejected was expressed
420 by some of our respondents but the acceptance received from partners instilled
421 confidence and self-esteem.

422 Although some impact on activity and participation was described, due to reduced
423 strength, pain and grip function, the participants managed to compensate or overcome
424 many practical obstacles by using effective problem-solving skills. Furthermore, an
425 inner strength and support system facilitated effective problem- and emotion-based
426 coping, as described by other groups of patients with hand injuries [22,36].

427

428 **Methodological considerations**

429 In qualitative research, the findings are evaluated in terms of trustworthiness, which
430 includes establishing credibility, dependability, confirmability and transferability [20,7].
431 Purposive sampling was used to provide variation in age, gender and cause of
432 congenital thumb anomaly. The interviews varied in length and depth, but were rich in
433 detail. The last four interviews provided little new information which ensured saturation
434 of data [20]. To strengthen dependability, three authors independently read the text and
435 engaged in in-depth discussions to arrive at a reasonable interpretation. Representative
436 quotations from the transcribed text are given to make the interpretation of the text
437 visible to the reader. Constantly confirming and clarifying information during the

438 interviews ensured confirmability. All the researchers participated in the analysis in
439 order to reduce the risk of over interpretation of the results due to the authors pre-
440 understanding of the phenomena in focus. Furthermore, the method used consistently
441 throughout the research process was analysis focused on the text, limiting the risk of
442 predetermined interpretation. The transferability of the findings may be limited to
443 patients experiencing appearance related concerns because of diseases or trauma in the
444 upper extremity.

445

446 **Implications for provision of care**

447 This study shows that the impact of a congenital hand anomaly on daily life varies. This
448 applies both to dealing with practical challenges and emotional consequences. Access to
449 support and effective coping strategies for those affected is vital. It is important to note
450 that healthcare professionals need training in how to deal with appearance-related
451 concerns. A pathological approach that uses negative terminology, such as defect,
452 deformity, abnormality and disfigurement, may exacerbate the difficulties experienced
453 by those seeking help [33,37]. A normalising approach for patients who are visibly
454 different may instead empower them and promote adaptive behaviour in social
455 situations. Some of our respondents indicated a need for professional assistance when
456 dealing with emotional consequences. The opportunity to meet other patients and share
457 experiences as well as having access to informative leaflets that would be useful in
458 school were also mentioned. Maddern et al show that cognitive behavioural therapy
459 (CBT), including social skills and problem-solving strategies, represents an effective
460 therapy for children with appearance-related problems due to congenital abnormalities
461 such as cleft lip and palate, burns or other forms of trauma A reduction in the frequency

462 of teasing and in the degree of distress it caused both in the classroom and in the
463 playground was seen at a six-month follow-up [32].

464

465 **In conclusion**

466 This study allows a deeper understanding of how being born with a visibly different or
467 missing thumb (thumb hypoplasia/aplasia and thumb duplication) may influence daily
468 life. The findings emphasize the importance for healthcare professionals addressing
469 appearance-related concerns, which may cause long-term emotional distress and social
470 consequences.

471

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474

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488 **REFERENCES**

489

- 490 1. Ekblom AG, Laurell T and Arner M. Epidemiology of congenital upper limb
491 anomalies in 562 children born in 1997 to 2007: A total population study from
492 Stockholm, Sweden. *J Hand Surg* 2010;35:1742-54.
- 493 2. Aliu O, Netscher DT, Staines KG, Thornby J and Armenta A. A 5-year interval
494 evaluation of function after pollicization for congenital thumb aplasia using
495 multiple outcome measures. *Plast Reconstr Surg* 2008;122:198-205.
- 496 3. Netscher DT, Aliu O, Sandvall BK et al. Functional outcomes of children with
497 index pollicizations for thumb deficiency. *J Hand Surg Am* 2013;38:250-7.
- 498 4. Patel AU, Tonkin MA, Smith BJ, Alshehri AH and Lawson RD. Factors
499 affecting surgical results of Wassel type IV thumb duplications. *J Hand Surg Eur*
500 *Vol* 2013; 39:934-43.
- 501 5. Staines KG, Majzoub R, Thornby J and Netscher DT. Functional outcome for
502 children with thumb aplasia undergoing pollicization. *Plast Reconstr Surg*
503 2005;116:1314-23.
- 504 6. Franzblau LE, Chung KC, Carlozzi N, Chin AY, Nellans KW and Waljee JF.
505 Coping with congenital hand differences. *Plast Reconstr Surg* 2015;135:1067-
506 75.
- 507 7. Berg BL. *Qualitative research methods for the social sciences*. 5th Edn. Boston:
508 Pearson Education Inc, Allyn and Bacon, 2004;269-70.
- 509 8. Wassel HD. The results of surgery for polydactyly of the thumb. A review. *Clin*
510 *Orthop Rel Res* 1969;64:175-93.

- 511 9. Kozin SH. Upper-extremity congenital anomalies. *J Bone Joint Surg Am* Vol
512 2003;85: 1564-76.
- 513 10. Manske PR, McCarroll HR, Jr. and James M. Type III-A hypoplastic thumb. *J*
514 *Hand Surg* 1995;20:246-53.
- 515 11. Kozin SH. Pollicization: The concept, technical details, and outcome. *Clin*
516 *Orthop Surg* 2012;4:18-35.
- 517 12. Tonkin MA. Thumb duplication: Concepts and techniques. *Clin Orthop Surg*
518 2012;4:1-17.
- 519 13. Gummesson C, Ward MM and Atroshi I. The shortened disabilities of the arm,
520 shoulder and hand questionnaire (QuickDASH): Validity and reliability based
521 on responses within the full-length DASH. *BMC Musculoskelet Disord*
522 2006;7:44.
- 523 14. Eriksson M and Lindstrom B. Validity of Antonovsky's sense of coherence
524 scale: A systematic review. *J Epidemiol Community Health* 2005;59:460-6.
- 525 15. Sullivan M, Karlsson J and Ware JE, Jr. The Swedish SF-36 Health Survey-I.
526 Evaluation of data quality, scaling assumptions, reliability and construct validity
527 across general populations in Sweden. *Soc Sci Med* 1995;41:1349-58.
- 528 16. Carlsson I, Cederlund R, Hoglund P, Lundborg G and Rosen B. Hand injuries
529 and cold sensitivity: Reliability and validity of cold sensitivity questionnaires.
530 *Disabil Rehabil* 2008;30:1920-8.
- 531 17. Irwin MS, Gilbert SE, Terenghi G, Smith RW and Green CJ. Cold intolerance
532 following peripheral nerve injury. Natural history and factors predicting severity
533 of symptoms. *J Hand Surg [Br]* 1997;22:308-16.

- 534 18. Mathiowetz V, Kashman N, Volland G, Weber K, Dowe M and Rogers S. Grip
535 and pinch strength: Normative data for adults. Arch Phys Med Rehabil
536 1985;66:69-74.
- 537 19. Mathiowetz V, Weber K, Volland G and Kashman N. Reliability and validity of
538 grip and pinch strength evaluations. J Hand Surg [Am] 1984;9:222-6.
- 539 20. Graneheim UH and Lundman B. Qualitative content analysis in nursing
540 research: Concepts, procedures and measures to achieve trustworthiness. Nurse
541 Educ Today 2004;24:105-12.
- 542 21. Carlsson IK, Edberg AK, Wann-Hansson C. Hand-injured patients' experiences
543 of cold sensitivity and the consequences and adaptation for daily life: a
544 qualitative study. J Hand Ther. 2010;23:53-61.
- 545 22. Cederlund R, Thoren-Jonsson AL, Dahlin LB. Coping strategies in daily
546 occupations 3 months after a severe or major hand injury. Occup Ther Int.
547 2010;17:1-9.
- 548 23. Chemnitz A, Dahlin LB, Carlsson IK. Consequences and adaptation in daily life
549 - patients' experiences three decades after a nerve injury sustained in
550 adolescence. BMC Musculoskelet Disord. 2013;14:252.
- 551 24. Wijk U, Carlsson I. Forearm amputees' views of prosthesis use and sensory
552 feedback. J Hand Ther. 2015;28:269-77.
- 553 25. Dion K, Berscheid E and Walster E. What is beautiful is good. J Pers Soc
554 Psychol 1972;24:285-90.
- 555 26. Blakeney P, Meyer W 3rd, Robert R, Desai M, Wolf S and Herndon D. Long-
556 term psychosocial adaptation of children who survive burns involving 80% or
557 greater total body surface area. J Trauma 1998;44:625-32.

- 558 27. Rumsey N and Harcourt D. Body image and disfigurement: Issues and
559 interventions. *Body image* 2004;1:83-97.
- 560 28. Andersson GB, Gillberg C, Fernell E, Johansson M and Nachemson A. Children
561 with surgically corrected hand deformities and upper limb deficiencies: Self-
562 concept and psychological well-being. *J Hand Surg Eur Vol* 2011;36:795-801.
- 563 29. Ardon MS, Janssen WG, Hovius SE, Stam HJ and Selles RW. Low impact of
564 congenital hand differences on health-related quality of life. *Arch Phys Med
565 Rehabil* 2012;93:351-7.
- 566 30. Lukash FN. Children's art as a helpful index of anxiety and self-esteem with
567 plastic surgery. *Plast Reconstr Surg* 2002;109:1777-86.
- 568 31. Bradbury E. The psychological and social impact of disfigurement to the hand in
569 children and adolescents. *Dev Neurorehabil* 2007;10:143-8.
- 570 32. Maddern LH, Cadogan JC and Emerson MP. 'Outlook': A psychological service
571 for children with a different appearance. *Clin Child Psychol Psychiatry*
572 2006;11:431-43.
- 573 33. Egan K, Harcourt D, Rumsey N and Appearance Research Collaboration. A
574 qualitative study of the experiences of people who identify themselves as having
575 adjusted positively to a visible difference. *J Health Psychol* 2011;16:739-49.
- 576 34. Joachim G and Acorn S. Stigma of visible and invisible chronic conditions. *J
577 Adv Nurs* 2000;32:243-8.
- 578 35. Griffiths C, Williamson H and Rumsey N. The romantic experiences of
579 adolescents with a visible difference: Exploring concerns, protective factors and
580 support needs. *J Health Psychol* 2012;17:1053-64.

- 581 36. Gustafsson M, Persson LO and Amilon A. A qualitative study of coping in the
582 early stage of acute traumatic hand injury. *J Clin Nurs* 2002;11:594-602.
- 583 37. Murray CE, Kelley-Soderholm EL and Murray TL. Strengths, challenges, and
584 relational processes in families of children with congenital upper limb
585 differences. *Families, Systems & Health* 2007;25:276-92.
- 586

587 Table 1. Characteristics of participants

Participant	Gender	Age group (years)	Type of congenital hand problem
1	M	21-25	Thumb hypoplasia
2	M	26-30	Thumb duplication
3	F	36-40	Thumb duplication
4	F	21-25	Thumb duplication
5	F	21-25	Thumb duplication
6	M	21-25	Thumb duplication
7	F	26-30	Thumb duplication
8	M	16-20	Thumb duplication
9	M	26-30	Thumb hypoplasia
10	F	36-40	Thumb hypoplasia
11	M	26-30	Thumb duplication
12	F	16-20	Thumb hypoplasia
13	M	16-20	Thumb hypoplasia
14	F	51-55	Thumb hypoplasia
15	F	16-20	Thumb hypoplasia

588

589

590 Table 2. Characteristics of participants with congenital hand problems
 591 (n=15).

Outcome measures	Median (range)
Quick DASH	4.5 (0-55)
Sense of Coherence (SOC)	74 (48-85)
Physical Functioning*	95 (75-100)
Role Physical*	100 (0-100)
Bodily Pain*	84 (10-100)
General Health*	82 (25-100)
Vitality*	75 (0-100)
Social Functioning*	100 (38-100)
Role Emotional*	100 (0-100)
Mental Health*	84 (44-100)
Cold Intolerance Symptom Severity	15 (4-58)
Pain at rest**	0 (0-100)
Pain on motion without load**	0 (0-100)
Pain on load**	20 (0-100)
Grip function**	30 (0-80)
Fine motor skill**	20 (0-90)
Weakness**	30 (0-100)
Appearance of the hand**	60 (10-100)

Grip strength (Jamar dynamometer)***	28 (10-54)
Key pinch strength*** (n=14)	6.2 (1-12)
Pinch strength*** (n=13)	6 (0-14)

592 Values are in median (range)

593 * Subscales in Short Form -36 questionnaire [15]

594 ** Numeric rating scale range (0-100). 0 represents no problem, 100 represents worst possible problem

595 *** Average kg (range)[18,19]

596

597

598