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ORIGINAL PAPER

Progressive lengthening of short congenital forearm stump in children for prosthetic fitting

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Abstract
Purpose The sufficient length of congenital forearm stump is essential for prosthetic fitting. In our study we reviewed the results of a series of forearm stump lengthening, observed complications and their outcomes. We evaluated possibilities of combined technique to reduce or avoid problems and complications in forearm stump lengthening.
Methods We retrospectively reviewed 18 children who have undergone forearm stump lengthening. In all patients the forearm lengthening was performed by means of Ilizarov frame. Additional flexible intramedullary nailing (FIN) was applied in two cases.
Results The mean lengthening gain was 4.6 cm. The planned lengthening gain was obtained in all cases. The mean healing index (HI) was 34.1 days/cm. The most reduced HI was observed in two cases of combined technique (Ilizarov frame with FIN): 25.4 and 27.0 days/cm. Considering complications and outcomes the results were classified according to Lascombes: grade I—5 cases, IIa—10 cases, IIb—2 cases, IIIa—1 case. In the long term follow-up all patients used their prostheses fixed at the forearm stump with natural function of elbow joint.
Conclusion Forearm progressive lengthening in children with congenital transverse deficiency of the forearm is justified in order to facilitate prosthetic procedures and to preserve natural function of elbow joint. Sufficient lengthening can be achieved within one operation with a low rate of major complications. In our experience a repeated lengthening of forearm stump is not mandatory.

Keywords Congenital forearm stump lengthening

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Introduction

Congenital transverse deficiency is a failure of limb formation producing an amputation like stump. The congenital forearm stump presents a rare pathology [1]. Congenital amputation occurs mostly in the upper extremity in the forearm [2, 3]. The most common site is upper third that is why it is not always possible to obtain an ideal stump for prosthetic fit [4–6]. The sufficient length of forearm stump is essential for prosthetic fitting and to preserve natural function of elbow joint. In such situation, bone lengthening appears to be a solution in order to fix the prosthesis at forearm stump level [6–8]. In literature, the progressive lengthening is considered as an efficient method to increase the length of forearm stump and it does not require a tissue expander in selected patients [9–11]. However, previous reported series are very short and application of external device on the forearm stump still appears to be not simple [9–12]. Additional problems related to the procedure are revealed: delayed bone consolidation, necrosis of adherent skin covering the tip of the stump, protrusion of sharp bone ends through the skin, flexion contracture of elbow joint [4, 5, 9, 11–13].

The objective of our study was to review the results of a series of 18 forearm stump lengthening performed in order for better prosthetic fitting, to evaluate observed complications and their outcomes. We also estimated possibilities of combined technique and additional procedures performed simultaneously with applying of external device to reduce or avoid problems and complications in congenital short forearm stump lengthening.

Material and methods

We retrospectively reviewed our prospective database of 18 patients who have undergone limb lengthening and deformity correction from June 2000 to September 2010. During this period, 21 patients with congenital transverse deficiency of the forearm have been supervised. Inclusion criteria were the congenital forearm amputation at the level of the upper third, unsuccessful attempts to apply a prosthesis fixed at forearm stump and no other surgeries done earlier. The criteria of exclusion were: the forearm stump operated before and the age older than 16 years. In accordance with these criteria, 18 cases were chosen to be evaluated. The results of these consecutive cases of surgical treatment were analyzed.

All patients presented unilateral congenital forearm amputation. In one case (a girl, five years old) we observed unilateral upper and lower limb involvement: congenital forearm and tibia amputation. Male to female ratio was 1:1, mean age was 6.8 ± 0.4 years for girls and 7.7 ± 0.7 years for boys. There was no case with positive family history for congenital amputation. We did not reveal any malformation syndrome with forearm transverse deficiency in our series. The mean radiological length was 5.7 cm (3.9 to 8.2) for the ulna and 3.8 cm (2.3 to 4.5) for the radius. Segment length was measured using PACS system. The mean age at surgery was 8.3 ± 3.01 years (from 5 to 15). Before lengthening two patients had limited elbow extention (deficit was 20°), another three patients presented slight deficit of elbow flexion (10 to 15° in comparison to contralateral elbow). The strength of the upper limb muscles was evaluated according to the Lovett scale.

All patients have undergone forearm lengthening by means of Ilizarov frame. The application of the Ilizarov device was similar in all the patients. External fixator was applied using one full ring proximally and one full ring distally (Fig. 1). We inserted three to four wires at the level of each ring. The diameter of wire was 1.5 mm. Radius and ulna were transfixed with, at least, two wires at proximal and distal levels. It means, one wire in proximal and another in distal ring were inserted through both bones, the radius and ulna. It was not necessary to use olives because the wire disposition at the 90° angle was obtained in all the cases. The application of Ilizarov device was followed by percutaneous osteotomy of both bones performed by means of a narrow chisel via an incision of 5 mm long.

The flexible intramedullary nails of 2 mm diameter were used in combination with Ilizarov frame in two patients. The initial length of nails was longer than length of ulna and radius in order to provide nailing of the entire bone by the end of distraction period. In such situation protruding ends of elastic nails were embedding into the stump tissue during distraction period. In the first patient we applied two antegrade nails with distal ends outstanding over stump tip (Fig. 2). In the other case, the radial retrograde nail had protruding end into soft tissues of elbow joint region, the antegrade ulnar nail also had outstanding proximal bent end (Fig. 3). This technique provided protruding intramedullary nail end completely covered by soft tissues. The initial length of IM nails was calculated depending on the expected lengthening amount. In previous publications the combined technique (external fixator and flexible intramedullary nailing) proved to be efficient in forearm lengthening from the point of view of reduced period of external fixation and accelerated bone consolidation [14–16].

In order to avoid piercing of the skin covering stump tip by distal bone ends we performed two specific procedures. The first consisted of pulling up soft tissue reserve above stump tip in the moment of insertion of transfixing wires instead of making of soft tissue reserve between two rings. This procedure was performed in all the cases. Additionally, in five cases of wispy papery soft adherent tissues covering the sharp bone ends we performed, as the first step of surgery, a resection of 2–4 mm of the sharp bone ends through eccentric curved approach. The subperiosteal distal contact has been done simultaneously to obtain the distal radio-ulnar synostosis of sufficient surface of the flat bone ends. Distraction was initiated by the fifth to seventh day after surgery at the rate of 0.25 mm four times a day until the planned length was obtained. The distraction rate varied depending on bone regeneration.

Eight patients with elbow flexion contracture, which was developed during distraction period, required an additional full ring in the humerus with three wires. This ring was connected to the forearm device by an anterior threated rod with hinges placed in the axis of desired correction. The rod was distracted by 0.5 mm four times per day. In the beginning of fixation period this anterior rod, that allowed the motion of the elbow, was removed. The frame removal was performed after the complete bone consolidation.

All the cases were analyzed according to the assessment criteria: amount of lengthening, healing index (HI), complications and their outcomes according to Lascombes classification (Table 1) [17], use of prosthesis fixed at the forearm stump and natural function in the elbow joint in long-term follow up. The Lascombes classification takes into consideration expected and obtained lengthening amount, HI, function of adjacent joints in a year after frame removal, complications and their outcomes. The mean follow-up was 25.5 months.

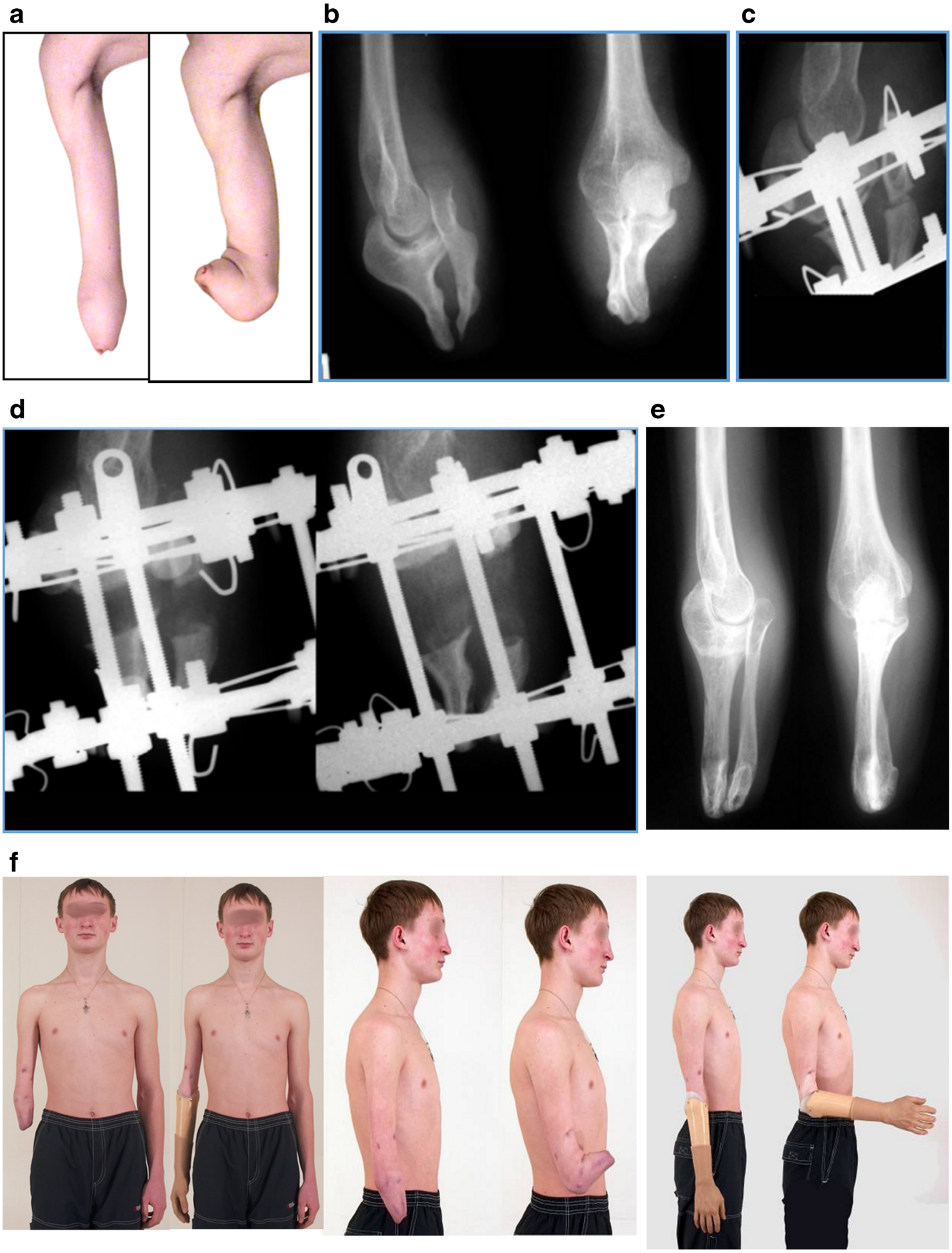

Fig. 1 Boy of 15 years with congenital forearm amputation: **a** upper limb before treatment; **b** radiographs of forearm stump before lengthening; **c** in the beginning of distraction period; **d** progressive lengthening of both bones of forearm stump; **e** radiographs of lengthened forearm stump with distal radio-ulnar synostosis; **f** functional outcome

Statistics were analyzed by means of the Attestat software (Attestation Software, Las Vegas, NV, USA). For the descriptive statistics, mean values of criteria and their standard deviation were defined.

Results

The mean distraction rate was 0.78 ± 0.17 mm/day. The mean achieved lengthening gain was 4.6 ± 0.47 cm (82.3 ± 18.5 %

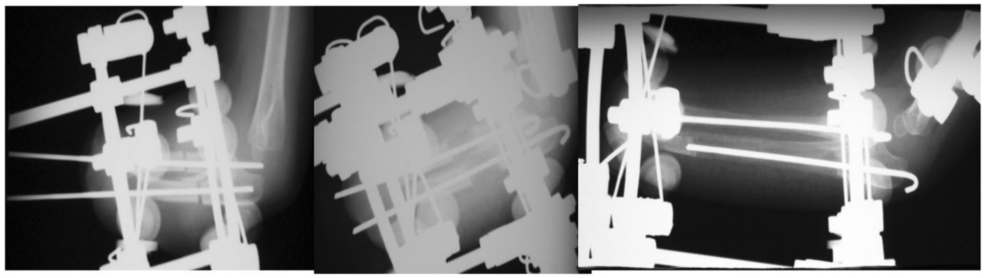

Fig. 2 Case of forearm stump lengthening with combined technique using antegrade initially protruding intramedullary nails

for the initial ulna length) (Fig. 1). The planned lengthening gain was obtained in all cases. The mean HI was 34.1 ± 5.37 days/cm. The most reduced HI was observed in two cases of combined technique (Ilizarov frame with FIN): 25.4 and 27.0 days/cm. The age at the moment of surgery, initial length of both bones, surgical technique, lengthening amount (in cm and % of initial length of ulna, HI) for each patient are presented in Table 2.

There were no major pin track infections requiring change of wires. On the other hand, we observed wires cutting skin in all the patients because of a very small initial gap between rings. The septic complications related to protruding FIN were not observed in both patients. In one case of delayed bone consolidation of lengthened ulna, the insertion of intramedullary hydroxyapatite flexible nail was performed in order to obtain bone consolidation (Fig. 4). One case of premature consolidation of the radius caused a loss of transosseous position of proximal wires. That is why the radial head was brought down. An unscheduled surgery was necessary and consisted of additional wires and reosteotomy to continue the distraction. The flexion contracture of the elbow joint appeared before frame removal to be a typical complication in forearm stump lengthening. There were eight cases of this contracture (with a loss of more than 50°) resisting to active kinesiotherapy. An unscheduled surgery consisted of applying of a humeral ring became necessary to reduce progressively the flexion contracture (Fig. 5). Only in two patients a slight flexion contracture (15° maximum) of elbow joint persisted in long term follow up. In one case we observed a transverse fracture at the level of humeral wires, which was treated by applying two supplementary Ilizarov rings. Only in one child the flexion contracture of elbow persisted in long-term follow up, which required biceps brachii tendon Z-plasty. The parents of this child refused to apply a humeral ring in distraction period when stiffness of elbow joint appeared and physiotherapy was not efficient. One patient manifested signs of protrusion of sharp bones through the skin by the end of first month after frame removal. The sharp bone end resection with local skin plasty was performed. Table 3 shows the list of complications, treatment, and evaluation of results according to the Lascombes classification [17].

In the long term follow-up of 25 months, on average, all patients were able to use their prostheses fixed to the forearm stump comfortably with natural function of elbow joint (Fig. 1). The strength of muscles was determined as very good (grade 5). The children used prosthesis employing their own elbow joint.

Discussion

Congenital amputation of the forearm is a rare abnormality, it is observed in 1/20000 live births [1]. Jain and Lakhtakia reported about 12 cases of congenital transverse deficiency through the upper third of the forearm among 612 cases of congenital orthopedic anomalies of a population of 3550 patients who attended the General Hospital in Godhra (India) for the purpose of obtaining a physically handicapped certificate [3].

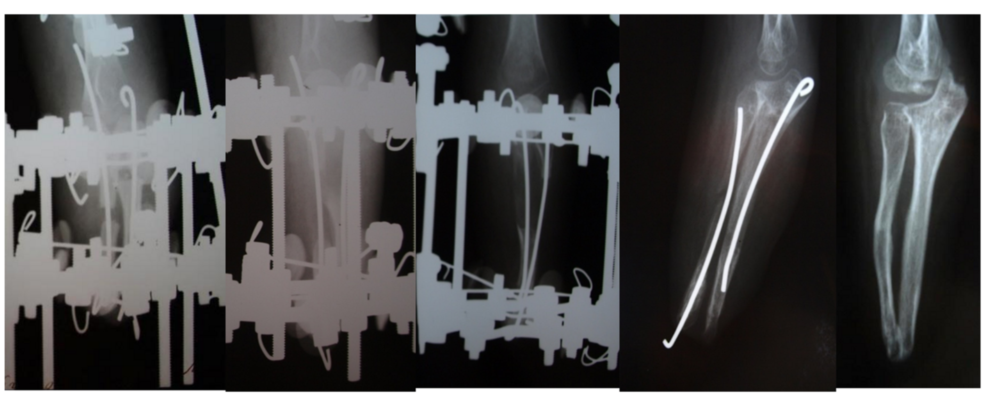

Fig. 3 Case of combined technique using antegrade ulnar and retrograde radial outstanding intramedullary nails covered by soft tissues

Table 1 Classification of results in limb lengthening (Lascombes et al. [17])

Grade	I	IIa	IIb	IIIa	IIIb	IVa	IVb
Additional surgery with general anesthesia	No	Yes, before frame removal	Yes, after frame removal	Possible	Possible	Possible	Possible
Lengthening gain obtained	Yes	Yes	Tes	Yes	No	Yes	No
Duration of treatment respected	Yes	Yes	Yes	No	No	Yes/No	Yes/No
Function respected	Yes	Yes	Yes	Yes	Yes	No	No

An available method of management of congenital below-elbow amputation stumps includes the fitting of prosthesis but requires an adequate length of stump [4–6]. In case of insufficient stump length an unsuccessful prosthetic outcome may be noticed almost in half of observations [18]. If the remaining stump is too short, a lengthening procedure may be beneficial [9, 10, 19].

Generally, there are three objectives for orthopaedic surgical treatment in children with congenital forearm transverse deficiency. First of all, a sufficient length of the stump is essential in order to apply a prosthesis at the forearm stump and to preserve natural function of the elbow joint [6, 9].

Second, sufficient soft tissue coverage of the stump tip to avoid ulceration of the skin and bone perforation [4, 5, 20].

Third, the child must have the necessary range of motion of elbow joint because even a functional forearm stump length is of no benefit in the case of adjacent joint contracture [6, 21].

Many promising results for stump lengthening with the Ilizarov frame were published [5, 9–11]. The majority of authors consider Ilizarov method as an efficient forearm stump lengthening technique, which does not require a preliminary tissue expender. Lengthening of both, the radius and the ulna, allows us to shape a rounded stump rather than a conical one for fitting of the prosthesis, but in selected patients [9]. The use of monolateral fixator is also possible in stump lengthening [8]. Segarra et al. proposed to lengthen forearm as the second step surgery after non-vascularized autograft of iliac crest with growth cartilage [12]. Milliez et al. used lengthening with Wagner device associated with groin flap resurfacing the distal end of forearm stump [22].

In literature, the achieved lengthening gain varies from 2 [5] to 14 cm [20]. The majority of authors concludes that amount of lengthening, 4 to 6 cm, is sufficient to have forearm stump length of 7–10 cm and to perform prosthesis fitting at the level of forearm stump [4, 5, 7, 9–11]. In our series, the mean lengthening of 4.6 cm (82.3 % of initial length of ulna, in average) allowed the prosthesis fitting and natural articulation in elbow joint in the mean follow-up of 25.5 months.

Duration of external fixation seems to be long in previous published studies: from four months to 217 days [5, 9–12]. In our series the mean HI was 34.1 days/cm but the combined technique of lengthening allowed to reduce it significantly: HI

Table 2 Age at the moment of surgery, initial length of both bones, surgical technique, lengthening amount (in cm and % of initial length of ulna), HI

Patient	age	Initial radius length; cm	Initial ulna length; cm	Technique of lengthening	Lengthening amount; cm	Lengthening amount; %	HI; days/cm
D.	5	4.0	6.4	Ilizarov	4.0	62.5	35.2
P.	5.5	2.8	4.1	Ilizarov	4.5	109.8	36.0
K.	9	2.3	3.9	Ilizarov	5.0	128.2	27.6
F.	12	4.2	6.3	Ilizarov + FIN	4.7	74.6	25.4
U.	15	3.7	5.1	Ilizarov	4.8	94.1	28.9
S.	14	4.5	8.2	Ilizarov + FIN	5.0	61	27.0
Ch.	6	3.5	4.2	Ilizarov	4.0	95.2	29.8
G.	6	3.0	5.5	Ilizarov	5.0	90.	32.5
D.	5	3.4	5.9	Ilizarov	3.9	66.1	34.8
H.	11	4.4	6.5	Ilizarov	4.6	70.8	37.2
L.	7	3.8	5.2	Ilizarov	4.7	90.4	47.1
K.	8	4.0	5.9	Ilizarov	3.5	59.3	36.6
D.	8	4.2	6.1	Ilizarov	5.3	86.9	38.3
E.	7	4.2	6.4	Ilizarov	4.8	75.0	39.4
O.	7	4.0	6.3	Ilizarov	4.5	71.4	35.7
P.	6	3.9	5.3	Ilizarov	5.0	94.3	29.8
L.	7	3.8	5.2	Ilizarov	4.5	86.5	33.5
H.	10	4.3	6.7	Ilizarov	4.3	64.2	38.1

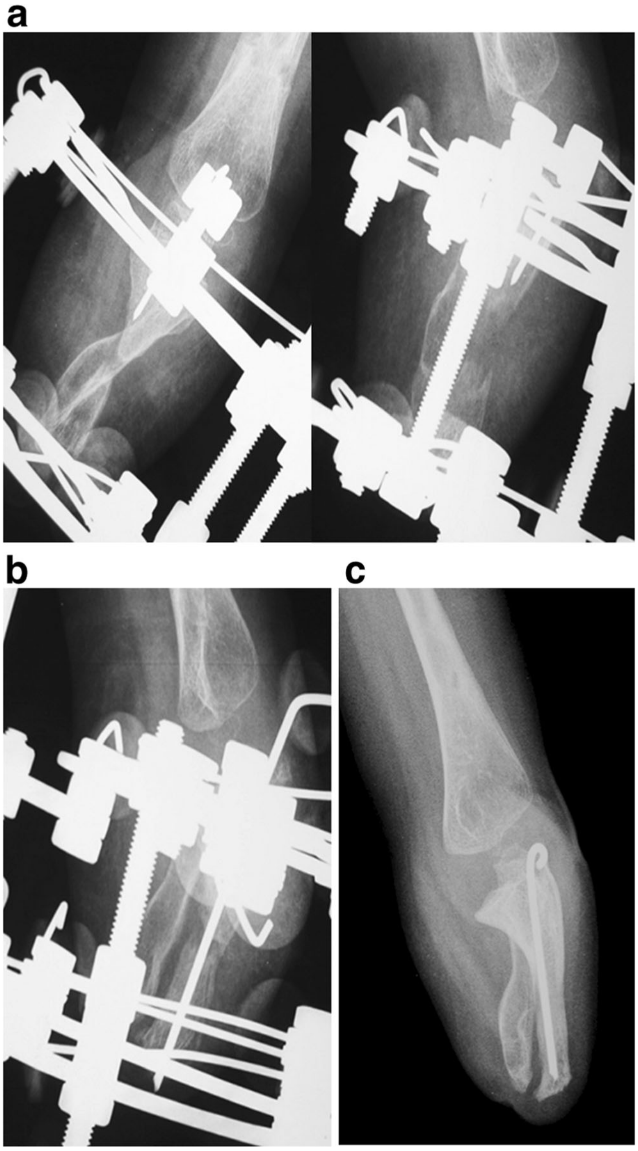

Fig. 4 Delayed consolidation of ulna: **a** weak bone regenerate with large soft tissue zone between two fragments of ulna; **b** insertion of hydroxyapatite-coated intramedullary nail; **c** bone consolidation

was inferior to 28 days/cm. This fact is in accordance with previous results in forearm lengthening with FIN [14–16]. The two cases demonstrate how to use the combined technique in bone lengthening in congenital short forearm stump.

Complications and additional procedures are common in forearm stump lengthening [4, 10, 11]. The temporary signs of pin track infection are similar in short forearm stump lengthening and do not influence the final result [9]. In our series all superficial infection around wires was cured locally and without any sequellae (grade I [17]).

The problem of flexion contracture of elbow joint is specific in forearm stump lengthening [4]. Jasiewicz et al. observed flexion contracture of elbow occurred during distraction period requiring an extensive kinesiotherapy program, Alekberov et al. found flexion elbow contracture in two patients of in a series of six cases [9, 11]. That stiffness is explained by the osteotomy site located more proximally than biceps tendon attachment due to very short congenital forearm stump. The Ilizarov method enables simultaneous correction of flexion contracture, but only if a humeral ring is applied. We prefer to use it only in cases of inefficient physiotherapy. In our patients the gradual distraction always allowed us to correct the flexion position of forearm, except one case of parental refuse of humeral ring. In such situation the Z-plasty of biceps tendon became necessary after frame removal.

All authors recognize the importance of soft tissue coverage of stump tip for functional outcome. For Bernstein et al. soft tissue coverage seemed to be the main limiting factor to lengthening [10]. Stricker reported a case of forearm stump lengthening complicated with impending necrosis of adherent skin covering the sump tip [13]. Different types of soft tissue plasty and flaps were proposed. Kour et al. used a flap with deep fascia and muscles components contoured around a stump simultaneously with bone lengthening by Ilizarov frame [20]. Segarra et al. had multiple complications in a short series, including problems of soft tissue covering stump tip [12]. Milliez et al. performed a groin flap resurfacing the distal stump after stump lengthening by Wagner device [22]. Alekberov et al., Orhun et al., and Jasiewicz et al. did not observe that kind of complication but in selected patients [5, 9, 11].

In our series we avoided protrusion of distal bone ends through stump tip during distraction period by use of the following procedures: resection of sharp bone ends, making soft tissue reserve above stump tip, distal radio-ulnar synosthosis. We emphasize the importance of performing these procedures simultaneously with applying an external frame. Only in one case the resection on protruding sharp bone ends was performed after frame removal. This case has been considered as an error in treatment planning.

All children in our series were able to use prosthesis comfortably employing their own elbow joint at the most recent follow-up. In contrast to some publications [11, 12], we observed no case of mandatory repeated lengthening. The majority of authors achieved sufficient functional length of stump with only one procedure [4, 5, 9, 20].

Our study is limited because of the small patient population. We cannot make a reliable conclusion on the influence of age of treatment on the outcome. According to Dabaghi-Richerand et al. [6], better functional outcomes are found in those children with congenital forearm amputation who started using a prosthesis before 6 years of age. Davids et al. noted that initial prosthetic fitting prior to the age of 3 years was associated with improved outcome [18]. It is important to emphasize that our results were achieved in a highly specialized center and this fact may decrease the reproducibility of results.

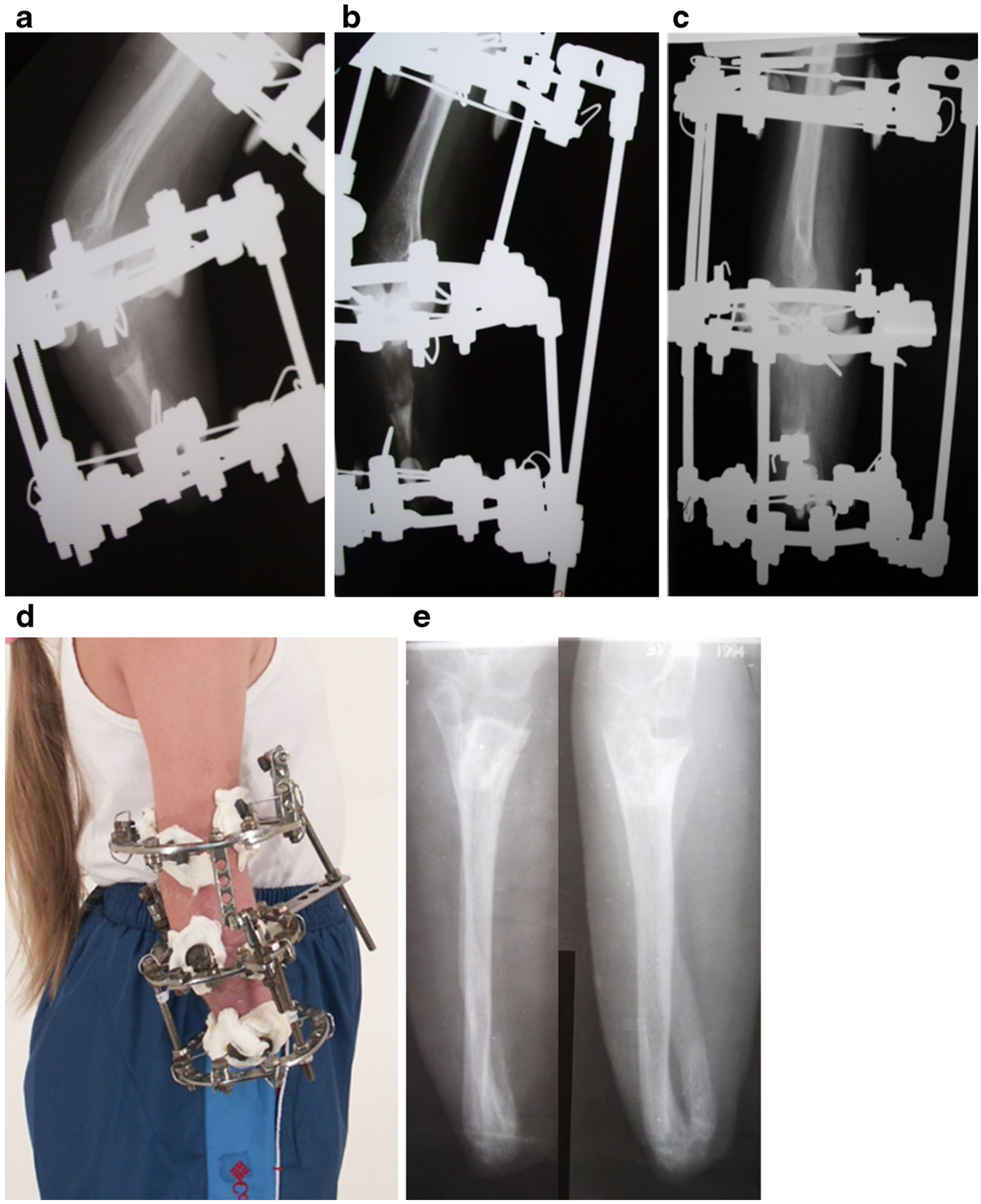

Fig. 5 Treatment of the flexion contracture developed during distraction period: **a** flexion position of forearm by the 14th day of distraction; **b** lengthening associated with progressive extension of elbow by means of anterior threaded rod; **c** planned lengthening amount of stump and complete extension of elbow are achieved; **d** aspect of external device for forearm stump lengthening with humeral ring and anterior rod; **e** after frame removal, lengthened stump with distal radio-ulnar synostosis

Table 3 Complications, treatment, and results

Complication (nb)	Nb (%)	Treatment	Grade Lascombes [17]
Local infection	12 (66.7 %)	Local care ± per os antibiotic therapy	I
Flexion contracture of elbow joint during distraction period	8 (44.4 %)	Humeral ring, progressive distraction between humeral ring and forearm device3	IIa
Flexion contracture of elbow joint in long term follow up	1 (5.6 %)	Z-plasty of biceps brachii tendon	IIb
Fracture of humerus at site of wires	1 (5.6 %)	Additional wires and rings	IIa
Premature consolidation of radius	1 (5.6 %)	Additional wires, reosteotomy	IIa
Protrusion of bone ends through stump tip skin	1 (5.6 %)	Resection of sharp bone ends	IIb
Delayed bone union (HI > 45 days/cm)	1 (5.6 %)	FIN with hydroxyapatite-coated nail	IIIa

Conclusion

Forearm progressive lengthening in children with congenital transverse deficiency of the forearm is justified in order to facilitate prosthetic procedures and to preserve natural function of elbow joint. Sufficient lengthening can be achieved within one surgery with a low rate of major complications. In our experience a repeated lengthening of forearm stump is not mandatory.

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Compliance with ethical standards

Conflicts of interest The authors have no competing interest to declare.

Informed consent Informed consent was obtained from all individual participants (parents or guardians) included in the study.

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