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SURGICAL TECHNIQUE FOR COMPLEX SYNDACTYLY IN APERT SYNDROME: A SERIAL CASE

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ABSTRACT

Introduction: Complex syndactyly in Apert syndrome, especially complicated with synonychia and synostosis, is a surgical challenge. The incidence of Apert Syndrome is reported to be approximately 1 per 100,000 to 160,000 live births and its incidence in Indonesia is not yet known. It is practically symmetrical causing significant dysfunction andinfection if not treated properly. The goals in the treatment are the separation of independent digits without disturbing function and growth, the creation of a lined commissure, the provision of skin cover for the denuded nail edge and exposed bone, and to creation of aesthetically pleasing individual fingertips with proper nails, nail folds, and adequatepulp fullness. This is due to the unreliability of the vascular branching pattern to the digits.
Case Illustration: In this serial case, we reported 5 cases of Apert syndrome. We described the clinical findings, incision design, immediate and post-surgery follow-ups.
Discussion: In this case, we combined dorsal rectangular flap and palmar triangular flap to make a new web space, Buck Gramcko pulp flap for the nail fold reconstruction, and full thickness skin graftw was taken from the groin to cover the raw areas. Absorbable 5.0 simple interrupted sutures were used in each patient. The results were uneventful, with satisfying function and aesthetic appearance.
Conclusion: Choosing the best surgical technique for releasing the web space in syndactyly depends on the patient's condition and the plastic surgeon's decision was needed. In 5 patients, up there was no meaningful complication attained from the postoperative result.

Highlights:

1. Complex syndactyly in Apert syndrome, particularly when complicated with synonychia and synostosis, poses a significant surgical challenge.
2. Successful surgical techniques for Apert syndrome syndactyly are crucial to achieving these goals.

INTRODUCTION

Apert syndrome is classified as type 1 Acrocephalosyndactyly is among the 5 types of acrocephalosyndactyly in craniosynostosis syndrome.[1] The first report was by Wheaton in 1894 and a French Paediatrician, Eugene Apert, and published a series of nine cases in 1906.[4] It Is a congenitaldisorder characterized by craniostenosis, exorbitism, midface hypo-plasia and symmetrical syndactyly of hand and feet.[1-7] Syndactily in Apert Syndrome is a complex anomaly of the bones, nerves, joints and the tendons.[1] It is commonly classified into 3 types that corresponds with the severity, the first type consists of a

complex (osseous and soft-tissue) syndactyly of digits 2 through 5, with the thumb largely free. This is described as the obstetrician's or spade hand.[1,2,6] The second type involves a complex syndactyly of digits 2 through 5 with an associated simple syndactyly of the thumb. It is also known as the spoon or mitten hand.[1-4,5] And the last type consists of complex syndactyly of digits 1 through 5, with complex syndactyly of the thumb. This type is type III (rosebud or hoof hand) that presents as a cup like hand. The functional prognosis of this type is uncertain.[1,2,6] In this serial case, we report 5 cases with complex syndactyly in Apert Syndrome and describe each case regarding its clinical findings and surgical management.

CASE ILLUSTRATION

Case 1

A two-year-old female diagnosed with Apert syndrome had type 1 syndactyly without any nail infection (Figure 1). The radiograph showed the bone of index-long-ring digits are joinedin both hands (Figure 2). It was decided that to do staged syndactyly release to avoid vascular compromise, prioritizing the release of the index fingers. For the second web space the incisiondesign was for a dorsal rectangular flap and a V-shaped palmar flap. To reconstruct the tips of the distal phalanges, buck-gramcko pulp flap design was made to recreate the nail fold (Figure 3). The shared nails were divided andthe united distal phalanges were separated by osteotomy. Remaining defects were covered with full thickness skin graft (FTSG) absorbable 5-0 suture was used to merge the skin (Figure 4). Pressure dressing and bandage and hand splints were used to immobilize the graft. On the 10th post-operative day, the outcome was assessed showing uneventful healing (Figure 5).

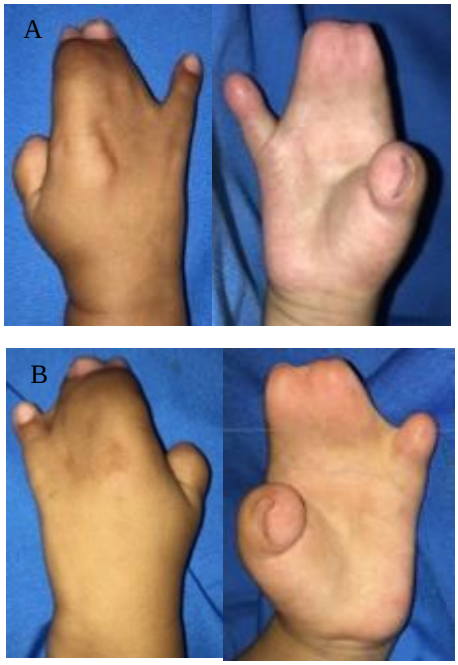


Figure 1. Presurgery Photo of Type 1 Syndactyly or Spade Hand, (A) Right hand. (B) Left Hand

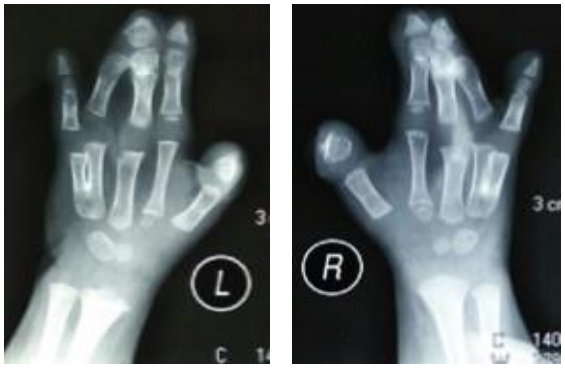
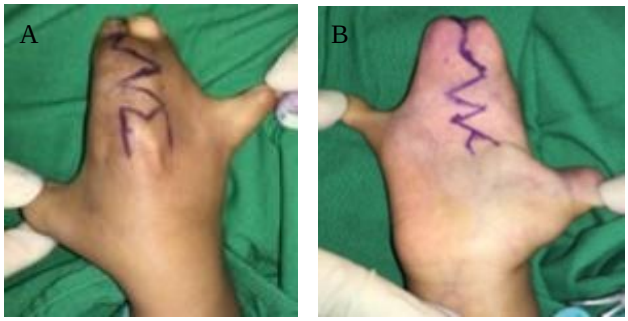


Figure 2. X-Rays of Left and Right Hands Showing The Extent of Bony Fusion



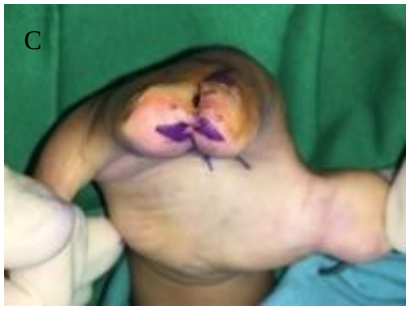


Figure 3. (A) Dorsal incision outline with a proximally based dorsal flap. (B) Palmar incision outline. (C) Buck-gramcko pulp flap design

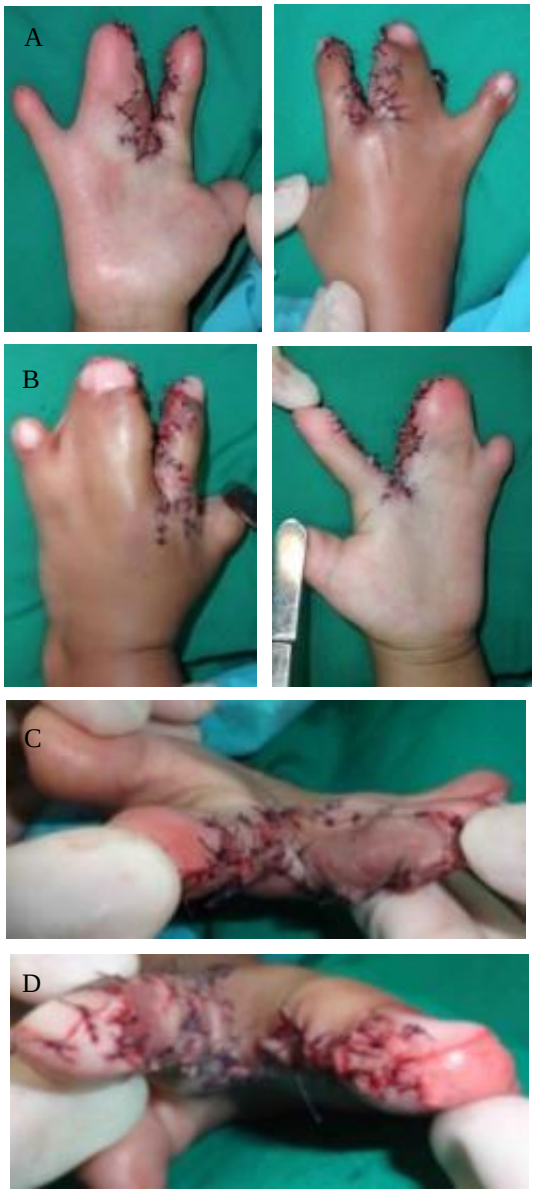


Figure 4. Immediate post-op photos, (A) Right hand (B) Left hand (C) The new webspace and FTSG coverage right hand (D) The new webspace and FTSG coverage left hand

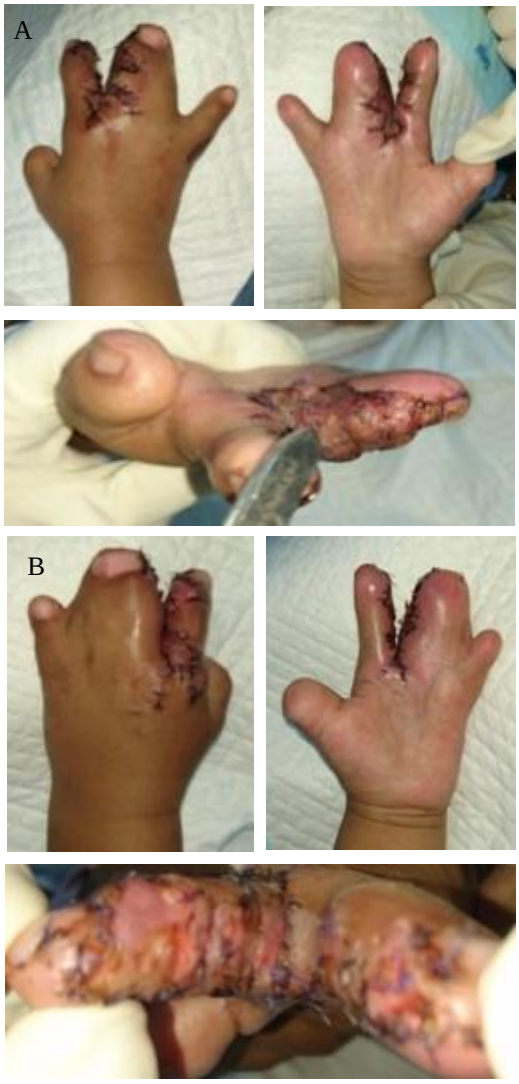
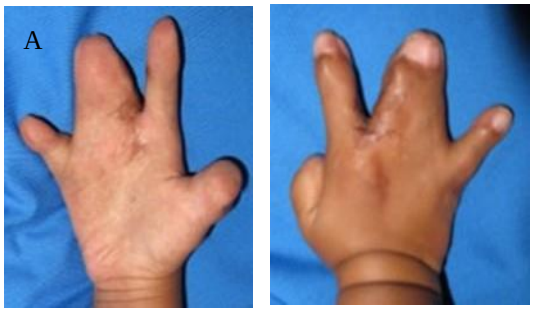


Figure 5. Follow up 10 days after surgery (A) Right hand. (B) Left hand



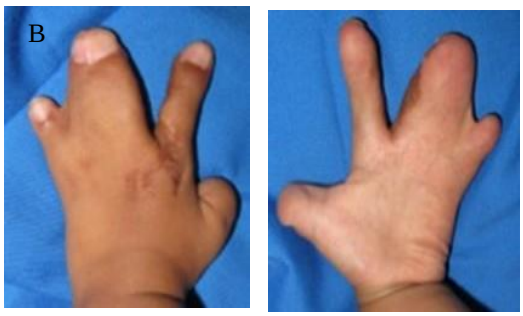


Figure 6. Follow up 6 months after surgery

Case 2

Female 19 months old presented with type 2 syndactyly or mitten hand. (Figure 7). Hand imaging showed bony union of the index- long-ring-little digits from the distal through the proximal phalanges and simple syndactylyof the first webspace (Figure 8). It was decided tofirst release the first web space bilaterally, and at the same time to also release. The second web space is on the left hand (Figure 9). An absorbable 5-0 suture was used (Figure10). Follow-ups to the 4th month after surgery showed uneventful healing (Figure 11).

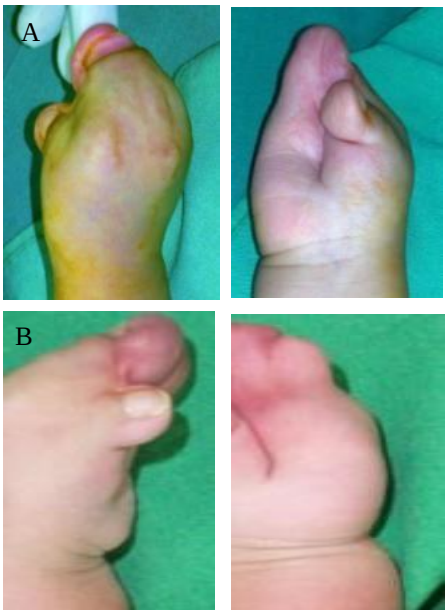


Figure 7. Pre-surgery photo of Type 2 syndactyly. (A) Right hand (B) Left hand

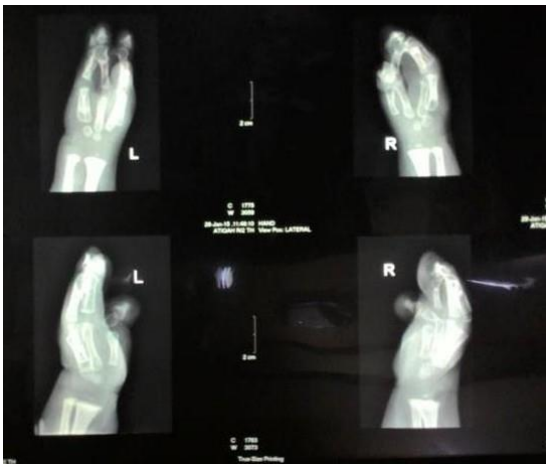


Figure 8. Pre-surgery hand radiograph of both handsshowing the extent of bony fusion

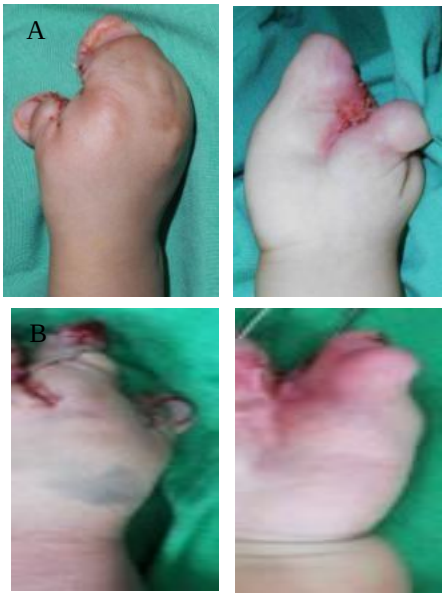


Figure 9. Intraoperative photos (A) Right hand (B) Left hand

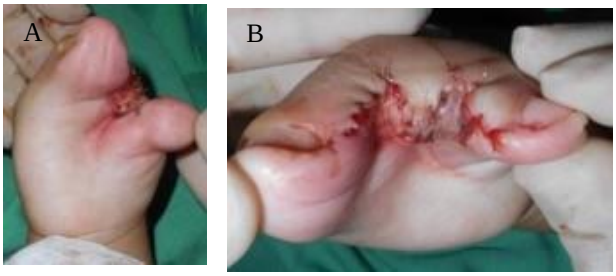


Figure 10. Post-surgery photos (A) Right hand (B) Left hand

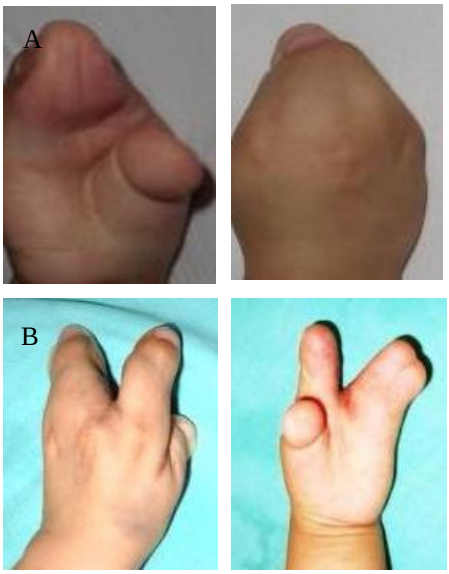


Figure 11. Follow up 4 months after surgery (A) Right hand (B) Left hand

Case 3

Male, 2 years old, presented with type 3 syndactyly or rosebud hand (Figure 12). Hand x-ray showed the bones of thumb-index-long-ring-little digits are joined at the distal, middle, and proximal phalanges. (Figure 13) Multiple V-shaped interdigital flaps and Buck-gramcko pulp flaps were also used on this patient (Figure 14). In the first stage, the thumbs were separated and re-aligned then the first web spaces were reconstructed. Follow-up at 7 days after surgery showed no vascular compromise of the flaps (Figure 15).

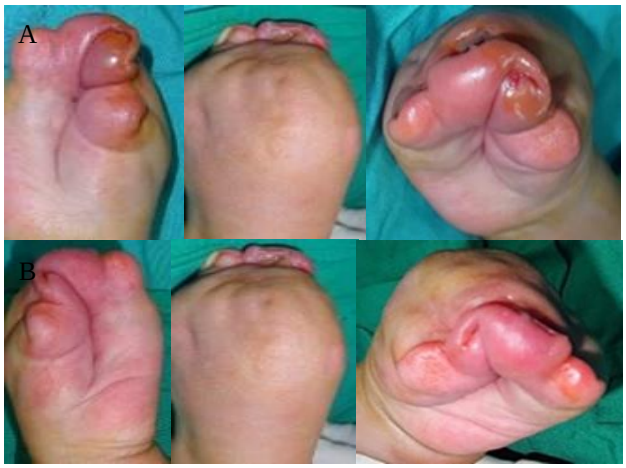


Figure 12. Type 3 syndactyly before surgery, (A) Right hand (B) Left hand

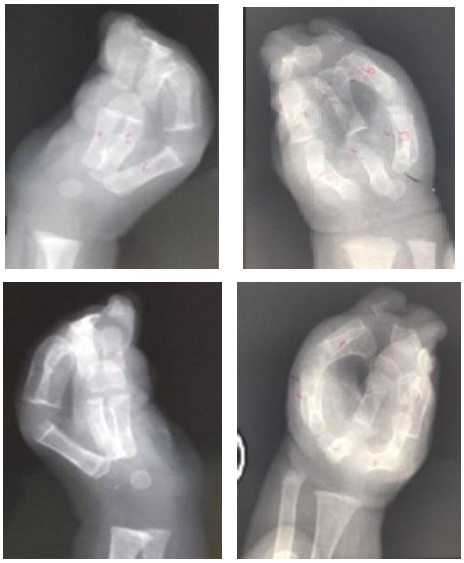


Figure 13. Hand X-ray before thumb release, showing the extent of bony fusion

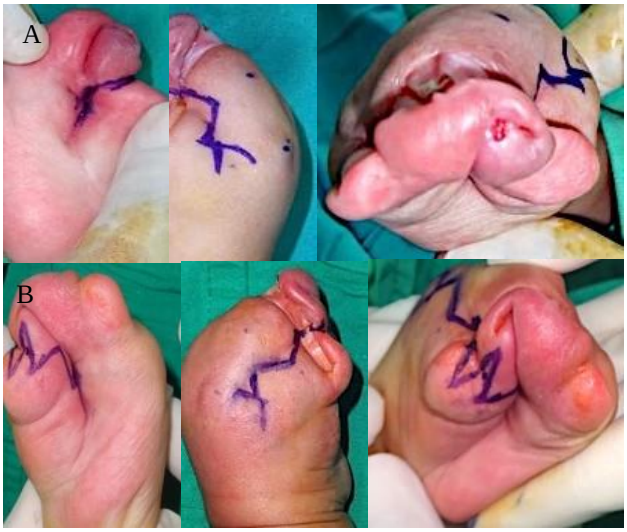
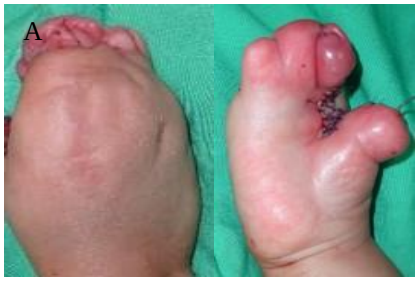


Figure 14. Surgical Design to release thumb (A) Right hand (B) Left hand



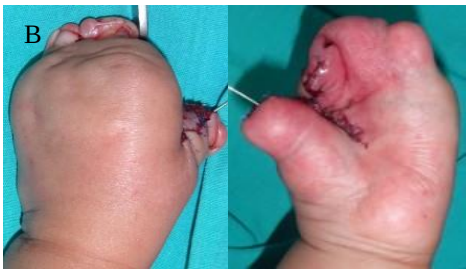


Figure 15. 7 days after first surgery (A) Right hand (B) Left hand

In the second stage (7 months later) the third web spaces were reconstructed after the third and fourth digits were separated (Figure 16). The hand X-rays after the 1st operation is are shown (Figure 17). Proximally based dorsal rectangular flaps and V-shaped palmar flaps were used to reconstruct the third web spaces and these extend from the metacarpal heads to two-thirds the length of the proximal phalanges. A zigzag incision to form interdigital flaps is then used to separate the digits. Another zigzag incision mirroring the dorsal incision was done on the palmar side. The nail fold was created using Buck-Gramcko pulp flap (Figure 18). Full-thickness skin graft were used to cover the remaining defects (Fig. 19). Photos 5 days after the surgery showed no complications (Figure 20).

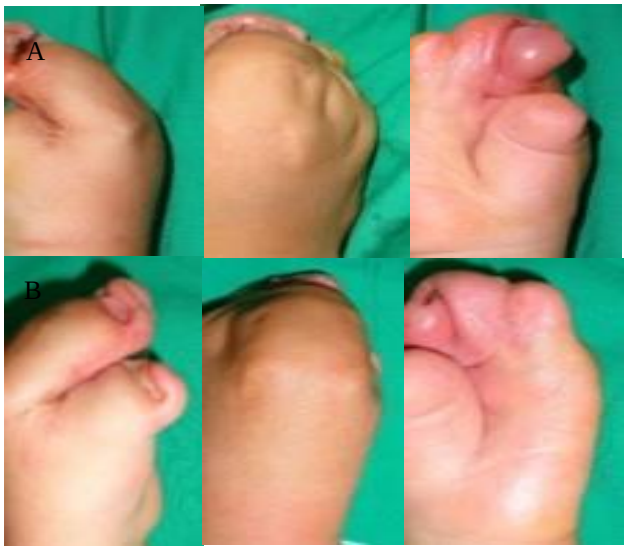


Figure 16. Before 2nd surgery (A) Right hand (B) Left hand

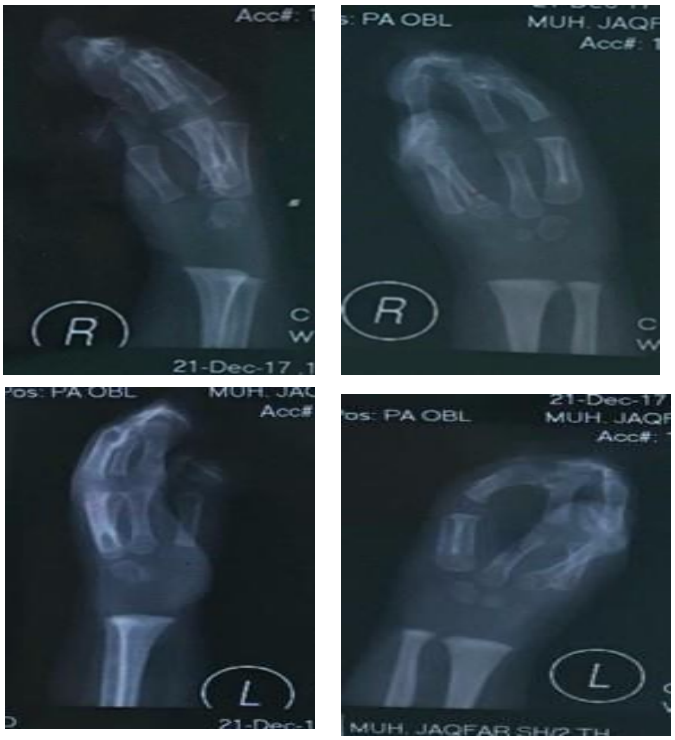


Figure 17. Pre 2nd surgery hand radiograph of both hand showing the extent of bony fusion of the

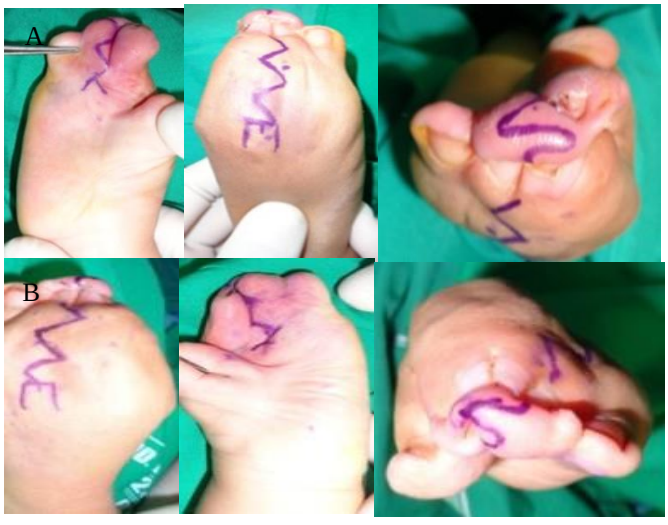


Figure 18. The nail fold was created using Buck-Gramcko pulp flap (A) Right hand (B) Left hand

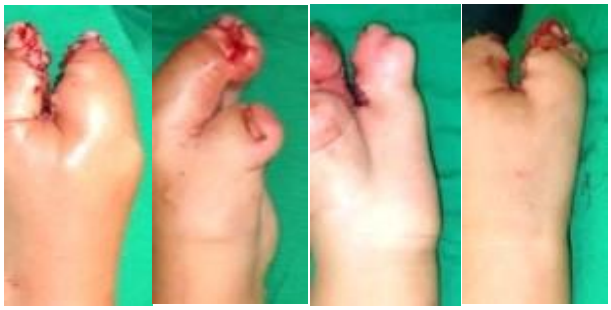


Figure 19. Immediate post-op photos

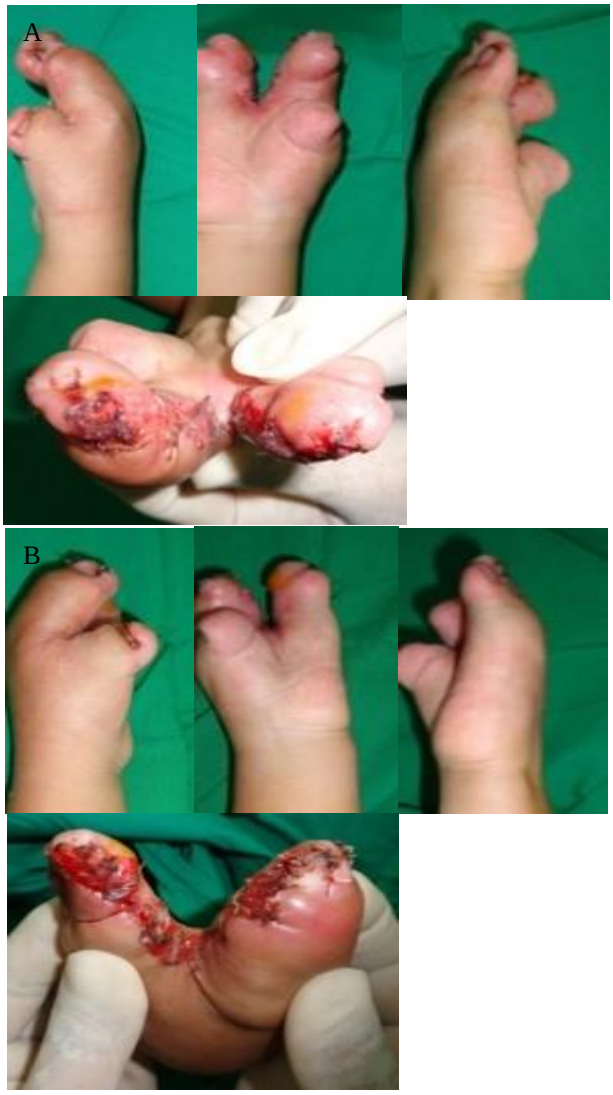


Figure 20. Follow up 5th days after surgery (A) Right hand (B) Left hand

Case 4

Male 4 years old diagnosed with Apert syndrome had type 3 syndactyly or rosebud hand (Figure 21). Hand X-rays shows the thumb-index-long-ring-little digits are joined at both hands (Figure 22). Dorsal palmar flap and Zigzagincision was design to use in this patient to release the thumb, remaining the defects were covered with full thickness skin graftFTSG (Figure 23).

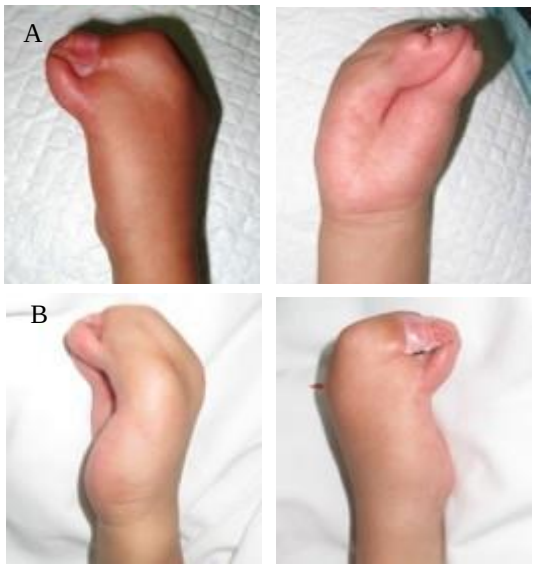


Figure 21. Clinical finding (A) Right hand (B) Left hand

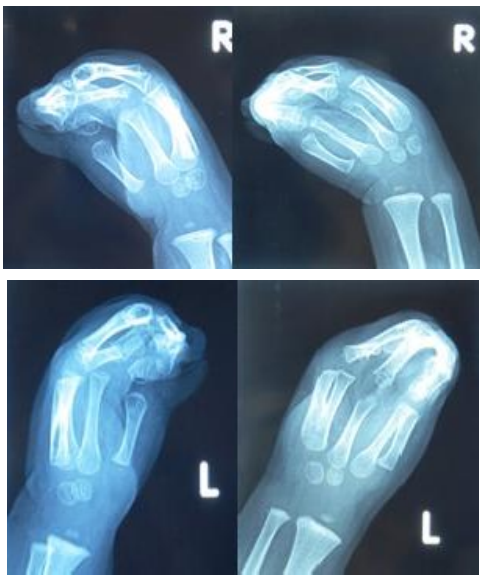


Figure 22. X-ray of both hand showing the extent of bony fusion

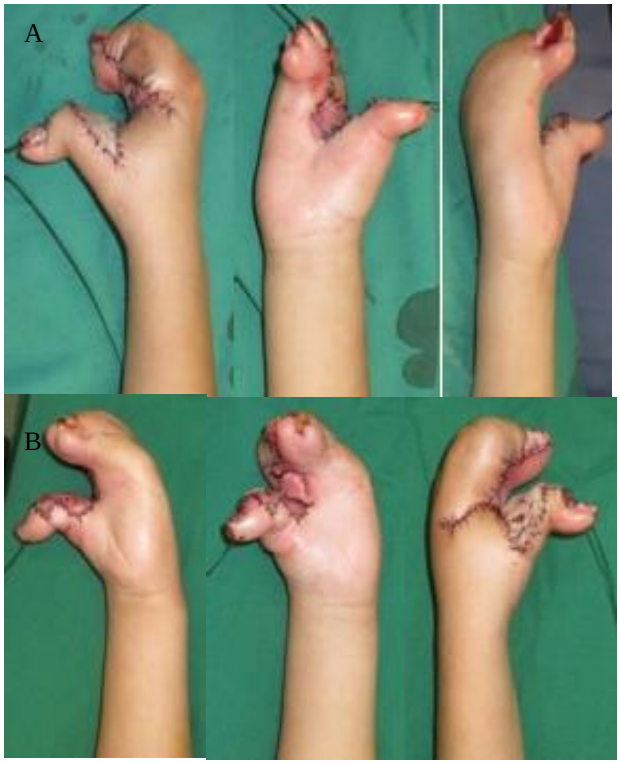
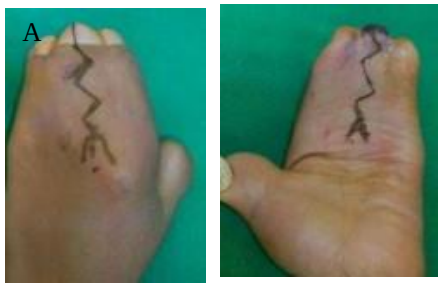


Figure 23. Early after surgery photos (A) Right hand (B) Left hand

Case 5

Male 6 years old presented with Apert syndrome and first type syndactyly or spade hand (Figure 24). X-ray showing the fusion distal phalanx of long-ring digits on the right hand and long-ring-little digits on left hand (Figure 25). This patient was also treated using dorsal rectangular flap and a V-shaped palmar. A zigzag incision is used to separate the web of the digits on dorsal and palmar is mirroring from the dorsal. We performed two web release at the right hand (2nd and 4th web) and 3rd web release on the left hand. (Figure 26) During the surgery the both hand has no significant complication.



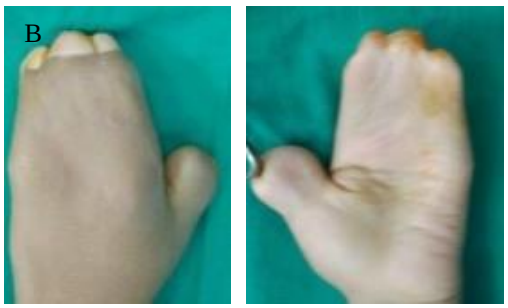


Figure 24. Type 1 syndactyly (A) Right hand (B) Left hand

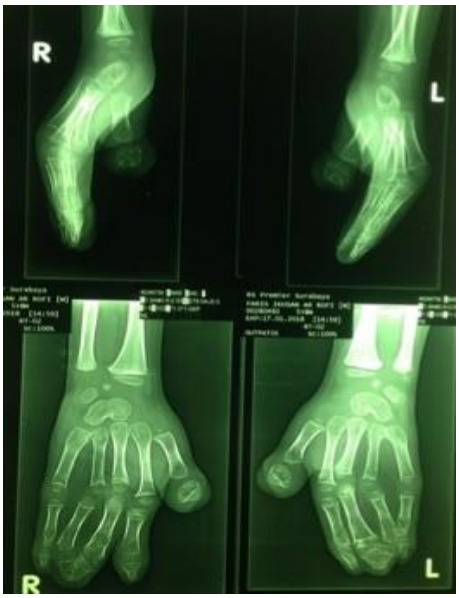


Figure 25. Pre-surgery X-ray of right and left hand showing the bony fusion

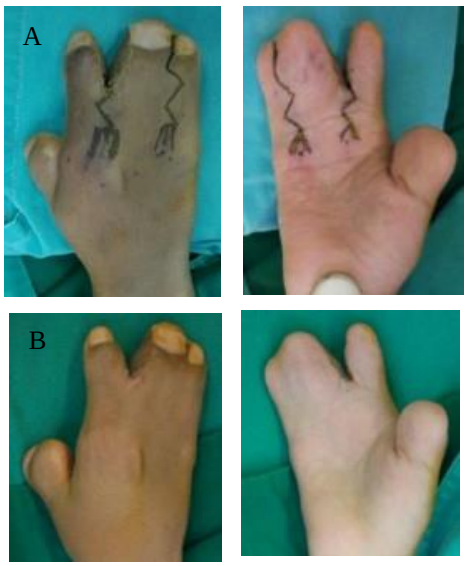


Figure 26. Surgical design to relase the web (A) Right hand (B) Left hand

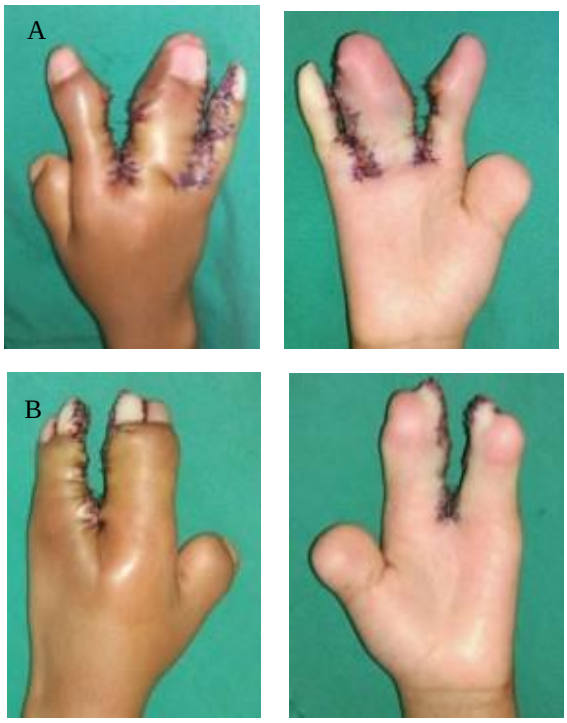


Figure 27. Early postoperative photos (A) Right hand (B) Left hand

DISCUSSION

Apert syndrome, also known as acrocephalosyndactyly syndrome type 1, is a rare genetic condition. It is characterized by the premature fusion of cranial sutures (craniosynostosis), severe syndactyly of both the hands and feet (fusion of fingers and toes), and distinctive facial features. This condition follows an autosomal dominant pattern of inheritance and is associated with mutations in the fibroblast growth factor receptor gene[12,14,15].

Almost all affected individuals experience craniosynostosis involving the coronal suture, with many also having sagittal and lambdoid suture involvement. In Apert syndrome, hand abnormalities consistently involve fusion of the middle three fingers, with potential involvement of the thumb and fifth finger in some cases[13].

The diagnosis of Apert syndrome is confirmed in an individual exhibiting classic clinical features (including multisuture craniosynostosis, midface retrusion, and syndactyly) and/or through the detection of a heterozygous pathogenic mutation in the FGFR2 gene using molecular genetic testing, alongside phenotypic characteristics that align with Apert syndrome[13].

The syndactyly in Apert syndrome is one of the most challenging problems due to vast involvement of soft and hard tissue of the hand. There are many literatures which emphasizes many aspects in the surgical management of Apert hand syndrome. The primary goals on syndactyly operations arethe capability to move the fingers freely in space, normal finger sensations and adequate covering, and also the ability to grasp objects with adequate power. There best period for syndactyly repair is 6 until 12 months of age. Reconstruction within this period will bring a maximum function and aesthetic outcome. Most of out patients were repaired after these period because of sosio-economic conditions.

There are a lot of additional principles that may be applied for the syndromic syndactyly. Most guidelines suggest to perform the surgical management in several stages to assure the operation goals are achieved. One of the pre-surgery assessment in complex syndactyly is computed tomography (CT) angiograms, it can visualization of the arterial anatomy of thehand so the surgeon can choose single or multiple release of the web. In our institution, didn't perform the CT angiograms so we released the web in multiple operations.

Several techniques that are commonly used in the Apert hand surgery are: Dorsalrectangular flap, V shape palmar

flaps, local rotation flaps, dorsal VY plasty, four flap z plasty, zig zag incisions, longitudinal incisions, free tissue transfers, Buck Gramcko's for nail fold reconstruction and combined with free flaps or a full-thickness skin graft are usually recommended to ensure adequate covering. External or internal fixation is sometimes used to guarantee the digit realignment. The techniques are plenty and are not perfect for one patient. Therefor the decision is tailored personally for each patient.

In our serial case, we combine dorsal rectangular flap for and palmar triangular flap are to make a new web space, Buck Gramcko pulp flap for the nail fold reconstruction, and full thickness skin graftwere taken from the groin to cover the raw areas. Absorbable 5.0 simple interrupted sutures were used in each patient. Meticulous care was given for the wounds. Sterile soft gauze and soft cotton elastic band are used to compress the wounds to immobilize the skin grafts. Dressing is maintained for 3 until 5 days and wound evaluation is performed.

From the follow ups, we evaluate short term and long-term complication. Most common short-term complications are wound infection, maceration of the flap or grafts, graft failure. Short term complications might be caused by seromas, hematomas, and inadequate immobilization. Long term complication might be seen after quite some time, which includes: web creep, hypertrophic scars, joint instability, hyperpigmentation and hair growth (from groin grafts), contractures, and nail deformities. All this patient in this serial case, has no serious complication at short or long term follow up.

The strength of this report are the surgical management of syndactyly in Apert syndrome, covering various aspects of the procedure and considerations. It clearly outlines the primary goals of syndactyly operations, which are the ability to move fingers freely, normal sensations, adequate covering, and the ability to grasp objects with adequate power. It emphasizes the importance of performing syndactyly repair between 6 and 12 months of age for maximum function and aesthetic outcomes. The paragraph mentions several surgical techniques commonly used in Apert hand surgery, providing insights into the approaches employed. It highlights the importance of tailoring surgical decisions to each patient's specific needs, recognizing the individuality of cases, and it discusses both short-term and long-term complications, offering a balanced perspective on potential outcomes.

It briefly mentions socioeconomic conditions as a reason for delayed surgery but does not delve into the potential impact of such constraints on patient outcomes, which could be relevant for discussion.

Overall, the paragraph provides valuable insights into the surgical management of syndactyly in Apert syndrome and a broader discussion of related factors and considerations.

CONCLUSION

The purpose of every principle technique is to obtain preferable outcome for normal digits, both in function and aesthetic. Choosing the surgical technique for releasing the web space in syndactyly depends on the patient condition and plastic surgeon's decision. In 5 patients that we had been followed up there was no meaningful complication attained from the postoperative result.

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CONFLICT OF INTEREST

The author declares that there is no conflict of interest in this case report.

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AUTHORS CONTRIBUTION

All authors contributed to the conceptualization, study design, data curation, formal analysis, data interpretation, methodology, manuscript writing, conceptualization, investigation, project administration, and content revision.

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