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A Comprehensive Clinical And Surgical Approach To The Treatment Of Complex Hand Syndactyly In Children With Apert Syndrome

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Abstract: Background: Apert syndrome is a rare congenital condition caused by mutations in the FGFR2 gene and is characterized by craniosynostosis, craniofacial deformities, and complex syndactyly of the hands and feet. Severe hand deformities lead to early functional impairment, limitation of self-care skills, and social adaptation difficulties in affected children. Aim. To evaluate the clinical and functional effectiveness of a comprehensive clinical and surgical approach in the treatment of complex hand syndactyly in children with Apert syndrome. Materials and Methods. Patients diagnosed with Apert syndrome and complex hand syndactyly underwent clinical observation. Evaluation included clinical examination to assess the severity of deformity, functional assessment of grasp and range of motion, and radiological examination. Treatment was carried out using a multidisciplinary approach. Surgical management was planned in a staged manner, taking into account skin deficiency, with the application of combined skin grafting techniques. Postoperative follow-up and rehabilitation were integral components of the treatment strategy. Results. Primary wound healing was achieved in the postoperative period. Adequate interdigital separation was obtained, and a significant improvement in hand mobility and grasping function was observed. The comprehensive clinical and surgical approach reduced the risk of postoperative

complications, including skin necrosis, contracture formation, and recurrence of syndactyly. Literature analysis supports the effectiveness of early and staged surgical intervention in improving functional outcomes in patients with Apert syndrome. Conclusion. A comprehensive clinical and surgical approach to the treatment of complex hand syndactyly in children with Apert syndrome provides favorable clinical and functional outcomes. Individualized surgical planning combined with postoperative rehabilitation significantly improves hand function and enhances the quality of life of affected children.

Keywords: IApert syndrome, acrocephalosyndactyly, complex syndactyly, symbrachydactyly, combined skin grafting, pediatric orthopedics.

1. Introduction: Apert syndrome (acrocephalosyndactyly type I) is a rare congenital genetic disorder associated with mutations in the FGFR2 gene and is characterized by premature fusion of the cranial sutures (craniosynostosis), distinctive craniofacial abnormalities, and complex syndactyly of the hands and feet. The estimated incidence of Apert syndrome ranges from 1:65,000 to 1:80,000 live births, making it one of the less common but clinically significant craniosynostosis syndromes.

The pathogenesis of Apert syndrome involves abnormal fibroblast growth factor receptor signaling, which leads to impaired differentiation and maturation of bone and soft tissues during embryonic development. As a result, patients often present with severe limb malformations, most notably complex syndactyly of the hands, characterized by fusion of both osseous and soft tissue structures. In many cases, this deformity manifests as symbrachydactyly, with shortened, fused digits and reduced interdigital spaces.

Hand deformities in children with Apert syndrome cause significant functional limitations from an early age, including impaired grasping ability, reduced range of motion, and difficulty performing fine motor tasks. These functional deficits restrict self-care activities and negatively affect psychosocial development and social integration. If left untreated or inadequately managed, complex syndactyly may lead to secondary complications such as joint stiffness, contracture formation, and recurrent deformities during growth.

Surgical correction of hand syndactyly in Apert syndrome represents a major challenge in pediatric orthopedic and hand surgery due to the severity of tissue fusion, associated skin deficiency, and the high risk of postoperative complications. Contemporary treatment strategies emphasize the importance of early intervention, individualized surgical planning, and staged reconstruction to minimize complications and optimize functional outcomes. A comprehensive clinical and surgical approach, incorporating multidisciplinary evaluation, combined skin grafting techniques, meticulous preservation of neurovascular structures, and structured postoperative rehabilitation, has become a cornerstone in the management of these complex cases.

Therefore, the development and evaluation of optimized clinical and surgical protocols for the treatment of complex hand syndactyly in children with Apert syndrome remain highly relevant, aiming to improve hand function, enhance quality of life, and facilitate long-term social adaptation of affected patients.

Aim of the Study

To evaluate the clinical and functional effectiveness of a comprehensive clinical and surgical approach in the surgical treatment of complex hand syndactyly in children with Apert syndrome.

2. Methods

This study involved the clinical observation of pediatric patients diagnosed with Apert syndrome accompanied by complex syndactyly of the hands. All patients underwent a comprehensive preoperative evaluation aimed at assessing the severity of anatomical deformities and functional impairment. Clinical examination included detailed inspection of the hands to determine the extent of syndactyly, the presence of bony and soft tissue fusion, skin deficiency, and the condition of the neurovascular structures. Functional assessment focused on evaluating grasping ability, finger mobility, and the range of motion of the interphalangeal and metacarpophalangeal joints, allowing for an objective estimation of hand function.

Radiological examination was performed using standard radiographs of the hands in multiple projections to assess skeletal abnormalities, the degree of osseous fusion, and joint alignment. Radiographic findings were used to support clinical data and to guide surgical planning. Based on the results of clinical, functional, and radiological assessments, each patient was discussed during a multidisciplinary consultation involving pediatric orthopedic surgeons, hand surgeons, anesthesiologists, and rehabilitation specialists.

The treatment strategy was developed individually for each patient, taking into account age, severity of deformity, functional limitations, and the availability of soft tissue coverage. Surgical intervention was planned and performed in a staged manner to reduce the risk of complications and to allow for gradual functional adaptation. Combined skin grafting techniques were applied to address skin deficiency and to achieve adequate interdigital coverage following syndactyly release. Postoperative management included regular follow-up examinations and a structured rehabilitation program aimed at restoring hand function and preventing secondary deformities.

Clinical Case Description

Diagnosis: Apert syndrome. Symbrachydactyly of digits II–V of both hands and digits I–V of both feet.

Surgical intervention: Stage I. Release of complex syndactyly between the II–III and IV–V digits of the right hand. Combined skin grafting was performed.

During surgery, the neurovascular bundles were preserved to the maximum extent possible, and anatomically adequate interdigital depth was achieved.

Figure 1. Preoperative and postoperative appearance of the right hand in a patient with Apert syndrome.

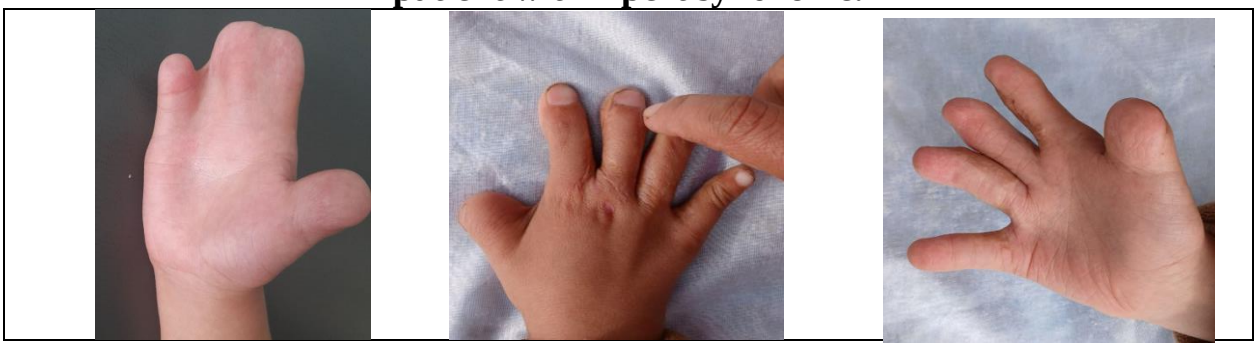
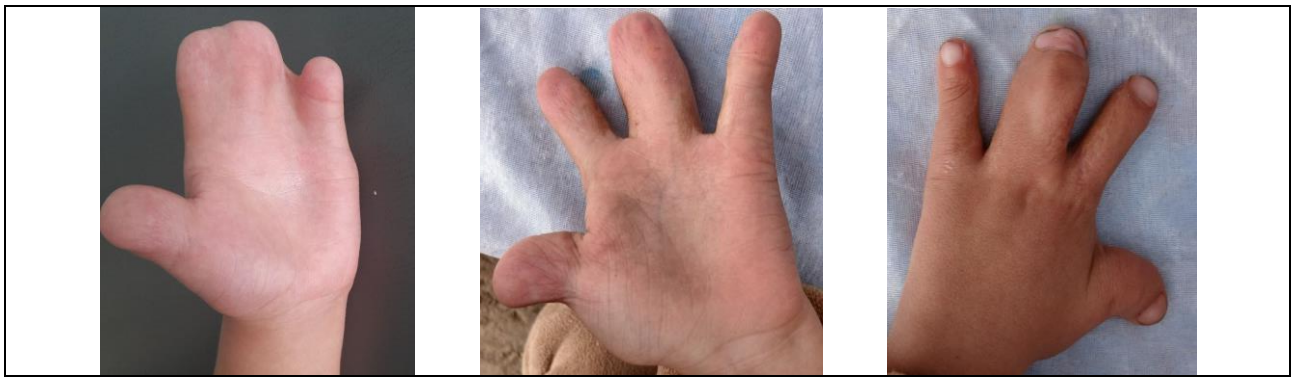


Table 1. Stages of Surgical Treatment of Hand Syndactyly in Apert Syndrome

Stage	Age	Surgical method
Stage I	2 years	Release of syndactyly between the II–III and IV–V digits of the right hand with combined skin grafting
Stage II	2 years 6 months	Release of syndactyly between the III–IV digits of the right hand with combined skin grafting
Stage III	3 years	Release of syndactyly between the II–III and IV–V digits of the left hand with combined skin grafting



Management Sequence for the Treatment of Complex Syndactyly in Apert Syndrome

The management of complex syndactyly in patients with Apert syndrome follows a structured and sequential approach aimed at achieving optimal anatomical and

functional outcomes. Treatment begins with a comprehensive clinical and radiological assessment to evaluate the extent of bony and soft tissue fusion, the severity of deformities, and functional impairment of the hand. Based on these findings, a multidisciplinary consultation is conducted involving pediatric orthopedic surgeons, hand surgeons, anesthesiologists, and rehabilitation specialists to determine the most appropriate treatment strategy.

Subsequently, an individualized surgical plan is developed for each patient, taking into account age, severity of syndactyly, skin deficiency, neurovascular anatomy, and functional demands. Surgical intervention is performed in a staged manner to minimize the risk of complications and to allow for gradual functional adaptation during growth. Postoperative management includes regular follow-up to monitor wound healing, finger alignment, and functional recovery, as well as early detection of potential complications. A structured rehabilitation program is an integral component of treatment, focusing on restoring hand function, improving range of motion, and preventing secondary contractures, thereby enhancing long-term functional outcomes and quality of life.

3. Results And Discussion

In the postoperative period, primary wound healing was achieved in all treated cases, with no signs of infection or significant wound complications. Adequate anatomical separation between the digits was successfully obtained, resulting in the formation of sufficiently deep and stable interdigital spaces. Preservation of the neurovascular bundles allowed for satisfactory tissue viability and contributed to favorable healing outcomes. Functional assessment demonstrated a noticeable improvement in the range of motion of the fingers, as well as enhanced grasping ability, which positively influenced the patients' capacity for self-care and daily activities.

The application of a comprehensive clinical and surgical approach, including staged surgical intervention and combined skin grafting techniques, significantly reduced the risk of common postoperative complications such as skin necrosis, scar-related contractures, and recurrence of syndactyly. Gradual correction through staged procedures allowed for better adaptation of soft tissues during growth and minimized excessive tension in the reconstructed interdigital areas.

Analysis of the available literature supports the findings of the present study, indicating that early surgical correction performed in a stepwise manner is associated with superior functional outcomes in children with Apert syndrome. Early intervention facilitates the development of hand function during critical periods of motor development and reduces the likelihood of secondary deformities. Multidisciplinary management and postoperative rehabilitation are emphasized in contemporary studies as essential components for achieving long-term functional success. Overall, the results confirm that an individualized, staged clinical and surgical strategy represents an effective and safe approach for the treatment of complex hand syndactyly in patients with Apert syndrome.

4. Conclusion

A comprehensive clinical and surgical approach to the treatment of complex hand syndactyly in children with Apert syndrome allows the achievement of stable and favorable clinical and functional outcomes. Early, staged, and individualized surgical planning, combined with meticulous preservation of neurovascular structures and the use of combined skin grafting techniques, plays a key role in reducing postoperative complications and improving hand function. Integration of structured postoperative rehabilitation further enhances functional recovery and contributes to a significant improvement in the quality of life and social adaptation of affected children.

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