The Apert Hand: Increasing Functional Outcome With Multistaged Syndactyly Release

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INTRODUCTION:

Apert Syndrome is characterised by craniosynostasis, facial hypoplasia and syndactyly with symphalangism of bilateral hands and feet.The classification and anatomy anomalies are well explained including normal features and variations present in Apert's hand [1].

Classification of the Apert hand can be divided into 3 types with the most complex form as type III or known as 'rosebud hand'. Controversies remain in the surgical treatment of Apert hand involving timing and number of operation,order of syndactyly release,types of skin closure and sacrification of finger [2]. Correction to increase functionality of the hands remains a difficult task especially in type III due to complex syndactyly,skin deficiency and overlapping of digits[3].

CASE DISCUSSION

We reported a 5 year-old girl with Apert syndrome both hands. She has complex syndactyly of bilateral hands , type III or rosebuds hand.

The child had two previous operation at the age of 2 years old and 6 months later. The aim was to convert a type III to a type I syndactyly where we created a first webspace between thumb and index finger and between ring and little finger. Creation of paronychial fold was done on second surgery.

The current third operation was done in view of contracture over webspace between left ring and little finger from the first operation. Zig zag incision was made at fourth webspace of left hand for contracture release. Vertical osteotomies done for right middle finger release. Again, full thickness skin graft was harvested for skin closure of the digits. Postoperatively, patient was able to perform pinch using right thumb, index and middle fingers.

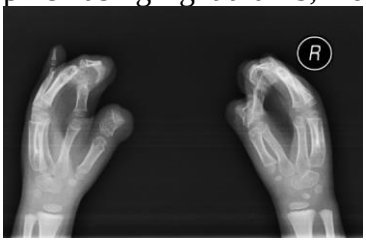

Figure 2

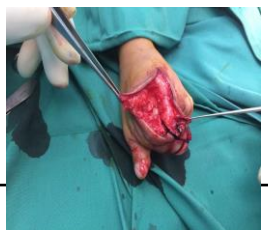

Figure 1: Radiograph showing synostosis between right proximal phalanx of index and middle fingers

Figure 2: Vertical osteotomies of synostosis between right proximal phalanx of index and middle fingers for middle finger release

DISCUSSIONS:

Apert syndrome is commonly associated with syndactyly of both hands and foot. Concerns were raised regarding corrective surgery in term of timing and number of operations, anesthetics challenge, pre operative planning, techniques of separation and type of skin closure[2].

The goal of release is before the patient begins schooling[3]. Upton et al recommends 2 stages of release with additional release if needed[5]. First surgery is to be done less than 6 months old. The aim is to create first and fourth webspaces with incision of nailfold for bilateral hands. A second surgery is done within 6 months later and/or before 3 years of age. The goal is for long ring release and re deepening of first webspace on unilateral hand. Additional surgeries can be made later when patient reaches preschool age for metacarpal synostosis, thumb clinodactyly correction and re deepening of web spaces. Principal of closure is reconstruction of the interdigital commissure with a local skin flap with/without additional full thickness skin graft.

CONCLUSION: Different techniques are available to correct complex syndactyly. The choice is usually made by the surgeon's preference and expertise. Nevertheless, the aim for surgery is to help patient to improve functional activities along with minimising complication from the surgery.

REFERENCES:

1. Harcourt, Patel, Kiran, Pradnya Sawant, and Dilip Chavan. "Anesthesia Management in a Patient of Apert Syndrome." *Anesthesia: Essays and Researches* 7.1 (2013): 133 London, pp 345–362

2. Upton J (2000) The Apert hand. In: Gupta A, Kay SPJ, Scheker LR (eds) The growing hand.

3. Sommerlad BC: The open finger technique for release of syndactyly, *J Hand Surg [Br]* 26:499-500, 2001.