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Surgical correction of wassel type 3 radial polydactyly: A case report

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Abstract
Radial or preaxial polydactyly is a common congenital anomaly occurring sporadically that must be treated early. Thumb duplications are a failure of formation and/or differentiation affecting the radial-ulnar axis of the hand plate in which Wassel has described seven types. The aim of surgical reconstruction is to obtain a stable and mobile thumb of adequate size and appropriate shape with preservation the function of pinch and opposition, and reconstruction of all components in one procedure. The most common form of reconstruction is removal of the lesser digit and reconstruction of the dominant digit. Here, we present a case of a Wassel Type 3 thumb duplication which was surgically corrected.

Keywords: duplication, thumb, radial polydactyly, wassel, surgical correction

Introduction
Congenital deformities of the hand are not uncommon and affects about 2.3 per 1,000 total births [1]. Thumb polydactyly is the most common duplication, especially in the Caucasian and Asian populations with an incidence of 0.8 to 1.4 per 1,000 births [2-4]. This condition is more a cosmetic rather than a functional problem, as the duplicated digit often contains all the sensory and motor units and that the duplicated thumb often works in unison with the whole hand as a single contributing unit [5]. Most patients with thumb duplication undergo excision of the duplicated thumb for either functional or cosmetic reasons.

Case report
9 year male presented to our department along with his mother with a deformity of the right thumb since birth. He was born to parents of second degree consanguineous marriage. He was born from a full term normal vaginal delivery with a good Apgar score. This deformity was identified immediately after birth. There were no other congenital deformities elsewhere. He was immunized appropriately for his age and he had normal developmental milestones till date. On clinical examination a diagnosis of a right radial polydactyly – Wassel type 3was made. X-ray of the hand confirmed the diagnosis. (Fig. 1) The child was able to write and hold objects, but the functions of the thumb were weaker compared to the opposite side. We planned for surgical correction of the deformity. Under general anaesthesia, tourniquet control and loupe magnification, a dorsal zig zag incision made and dorsal flaps elevated. Extensor pollicis longus (EPL) tendon and the radial collateral ligament were separated from the radial digit. The bone was isolated and an osteotomy was done at the bifurcation of the proximal phalanx and the radial split thumb was separated. EPL of duplicated digit was tagged to EPL of dominant thumb and the radial collateral ligament recreated in the thumb. Haemostasis secured and incision was closed in layers. Post-operative period was uneventful with a normal looking thumb with improved functions of pinch, grasp and opposition.

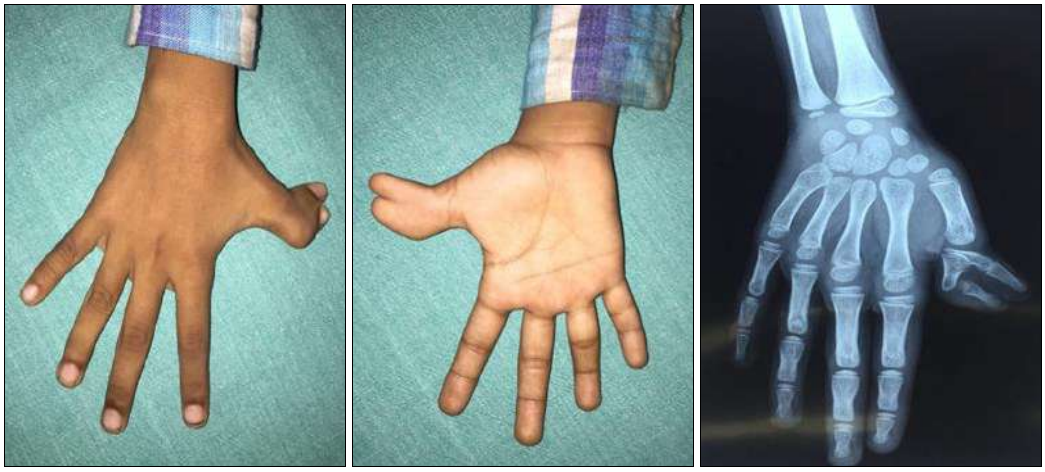

Fig 1: Clinical photographs showing the Wassel type 3 thumb with the X-ray confirmation

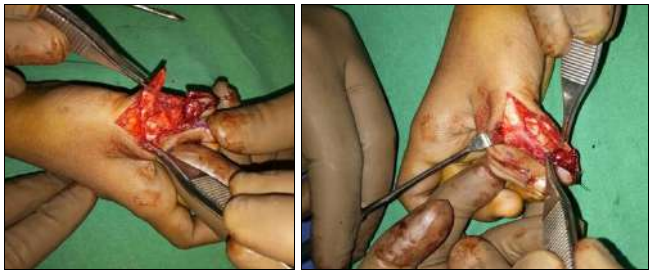

Fig 2: Surgical correction showing osteotomy of proximal phalanx and reconstruction of the radial collateral ligament

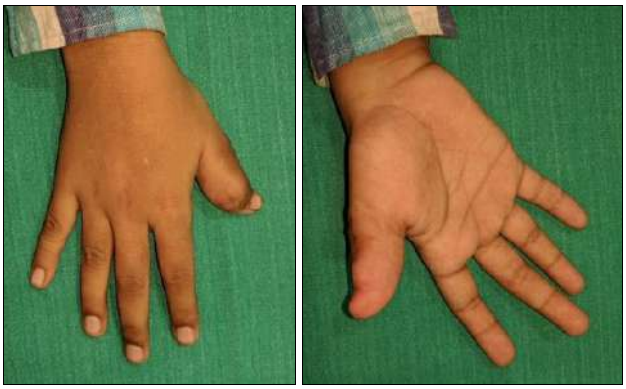

Fig 3: Late post-operative pictures showing a near normal looking thumb

Discussion

Thumb duplications result from a failure of the radial-ulnar axis of the hand plate to form and/or differentiate. The zone of polarizing activity (ZPA) in the posterior part of the developing limb bud is the principal signaling center. The ZPA expresses sonic hedgehog protein, which controls the formation of radial-ulnar features. Abnormal expression of Hox genes, bone morphogenic protein (BMP), and the Gli-3 gene all play a role in the evolution of thumb duplications [6]. Genetic counselling is not required as most cases are sporadic and unilateral. However, the presence of a triphalangeal thumb is known to exhibit an autosomal dominant inheritance pattern [4, 7]. Wassel in 1969 developed a classification system and it describes seven types of radial polydactyly according to the level of the skeleton at which duplication occurs [6, 8, 9]. Types I to VI classify distal-to-proximal levels of thumb duplication, and type VII describes radial polydactyly with a triphalangeal component. Type IV is the most common type of thumb polydactyly, followed by type II and type VII [10, 11]. In clinical practice, this is the most useful classification and it is used as a basic framework for the management of a thumb duplication [6]. A detailed evaluation is required in all patients with particular importance to thumb motion and functionality, status of the thenar musculature and first webs pace. Thumb stability and size relate to strength, both for grip and pinch whereas thumb mobility is largely dependent upon the integrity of the carpometacarpal (CMC) joint which is generally normal in Wassel types I to IV, may be slightly underdeveloped in types V and VI. Stability of the distal thumb is more important than motion, and if the CMC joint is intact, motion should be sacrificed for stability if necessary [6]. The flexor and extensor tendons of the thumb may also have eccentric insertions which can result in distal convergence of the duplicated thumbs [12]. The affected hand should always be compared with the opposite unaffected side as bilateral involvement could be syndromic, such as Townes-Brocks syndrome [12, 13]. X-rays are required to know the pattern of bone involvement and for follow-up as these are performed around one year of age, or at the time of surgery. X-rays performed prior to one year are not useful as there is a lack of ossification of the hand skeletal system [14]. Surgery is required for all cases of duplicated thumbs as the potential for improvement of function and appearance outweighs the risks of surgery. The aim of surgery is to achieve a stable and mobile thumb of appropriate size and shape with the restoration and maintenance of function and pinch activity [13]. Surgical correction must address the bony structural issues as well as soft tissue reconstruction including musculotendinous abnormalities which may encompass the radial and ulnar collateral ligaments and the flexor and extensor tendons for appropriate mechanical stability as simple ablative surgery results in instability and malalignment of the remaining thumb [4, 15]. Generally reconstruction is performed between 1 to 2 years as the pinch function is developing and before the development of fine motor skills [12]. The surgical options include resection and reconstruction (85%), simple ablation alone (5%), the Bilhaut-Cloquet procedure (8%), pollicization (1%), and on-top plasty (1%) [12].

Conclusion

Thumb duplication is a common congenital deformity encountered in infancy. The goal of treatment is to achieve a normal looking thumb with maintenance of function. The surgical corrective options of radial polydactyly are varied and complex and each subtype requires a specific surgical approach which determines the best overall result.

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