Gradual Ulna Lengthening And Elbow Reconstruction In A Patient With Multiple Hereditary Exostoses (MHE) With Negative Ulna Variance And Complete Radial Head Dislocation

¹Lim CH, ¹Nik Noraniza, ¹Kamariah Nor, ²Khairul Nizam Siron, ¹N Premchandran ¹Department of Orthopedics, Hospital Tengku Ampuan Afzan, 25100 Kuantan, Pahang ²Department of Orthopedics, Kulliyyah of Medicine, International Islamic University Malaysia, Kuantan, Pahang

INTRODUCTION:

Forearm deformity is common in patients with multiple hereditary exostoses where there is defective metaphyseal remodeling causing exostoses and growth retardation. The shortened ulna as result of early closure of distal physis caused radius to bow to accommodate, resulting in cubitus varus or radial head dislocation and limitations in forearm supination-pronation which can be aesthetically disturbing and functionally impaired.

CASE REPORT:

Ms. A is a 10-year-old girl with MHE who presented with worsening deformity of bilateral forearm and limitation in elbow and forearm movement as she ages which are disabling her. Our hypothesis was that patient would benefit from the gradual lengthening of ulnar using the Limb Reconstruction System (LRS) technique. However, during follow-up, we noted despite successful lengthening of ulna and deformity correction, the radial head is still subluxated due to the acute angle of radial neck resulting from chronic dislocation. We decided for elbow reconstruction with open radial head reduction, osteotomy and lateral collateral ligament reconstruction. Postoperatively, she was able almost entirely to supinate and pronate her forearm with no pain. Wrist and elbow range of movement was fairly good.

DISCUSSIONS:

Recent literature offers divergent perspective regarding treatment of these deformities. By using principles of distraction osteogenesis using the LRS, lengthening of ulnar with simultaneous correction of deformity became a surgical option. From a retrospective study by Yong and Sung with gradual ulnar lengthening with illizarof, there were good clinical and

radiological outcomes regarding radial bowing, ulnar shortening, carpal slip, and forearm pronation-supination. However, there was little change in preoperative radial articular angle and elbow flexion-extension arc.

Acute or gradual ulnar lengthening, distal radius epiphyseal stapling, radius corrective osteotomy or excision or open reduction may be included in corrective or reconstructive surgery of the forearm. However, there are no reports of the results of surgical treatment in MHE patients with dislocated radial head treated by gradual lengthening and elbow reconstruction. Symptomatic radial head instability appears to be an indication for ulnar lengthening surgery. Corrective radial osteotomy may be considered near the end of the growth spurt.

CONCLUSION:

There was no best method for treatment of MHE patients with forearm deformities. However, gradual lengthening of ulna with limb reconstruction system with or without elbow reconstruction is useful to treat forearm deformities by restoring the normal relationship of radius and ulna as closely as possible. Wrist and radial head stability, joint function and appearance are all improved.