

## Bilateral Proximal Radio-Ulnar Synostosis – Case Report

<sup>1</sup>Lee YK, <sup>2</sup>Cheah WN, <sup>1</sup>Ruzaimi MAR, <sup>1</sup>Zairuddin AZ, <sup>1</sup>Surianty S, <sup>2</sup>Noraini AR

<sup>1</sup>Department of Orthopaedic, Hospital Kuala Krai, 18000 Kuala Krai, Kelantan, Malaysia

<sup>2</sup>Department of Paediatric, Hospital Kuala Krai, 18000 Kuala Krai, Kelantan, Malaysia

### INTRODUCTION:

Congenital radioulnar synostosis is a rare forearm malformation causing restricted forearm supination-pronation in a various angles. The exact etiology is still not well understood but literature shown that it occurs in bilateral forearm for 60-80%, 9% run in family and 25% was genetically predisposed.

This report will highlight a young girl with soft dysmorphism features and was diagnosed to have proximal radio-ulnar synostosis after presented to us with restricted forearm range of motion.

### CASE REPORT:

Our case is a 2 year old young girl underlying Atria-Septal Defect (Secundum type) on anti-failure treatment, Left Eye Congenital Cataract and G-6-PD deficiency .She was born via vaginal delivery at 35 weeks of prematurity with birth weight of 2.2 kg.

She was brought by her mother due to bilateral forearm deformity with restricted movement. Otherwise, patient is an active young girl and there is no preceding trauma prior to the current abnormality.

On examination, she has soft dysmorphism features but not fulfill any specific syndrome. Local examination revealed her forearm is in fix pronation of left 70-80degree, right 30-40degree without passive or active supination. Her elbow x-ray (**Figure 1&2**) showed bilateral proximal radio-ulnar synostosis.

**Figure 1 Right Elbow X-ray:** Anteroposterior(right) and Lateral view(left) , an arrow showing visible osseous synostosis bridging between radius and ulna bone



**Figure 1 Left Elbow X-ray:** Anteroposterior(right) and Lateral view(left) , an arrow showing visible osseous synostosis bridging between radius and ulna bone



### DISCUSSIONS:

Congenital proximal radio-ulnar synostosis is a rare developmental deformity due to failure of longitudinal segmentation. The cartilaginous template bone was appeared from the upper limb bud as early as 37th days after fertilization. Initially, these cartilaginous template bones were connected together, especially for radius and ulnar bone that temporarily sharing a common perichondrium. Any factors disturbing its segmentation will cause bridging of the radius and ulnar bone. Eventually, it will lead to radius and ulnar synostosis in the form of osseous or fibrous connection.

Various classifications had been proposed based on the radiological features. Cleary and Omer et al [2] classification had divided the deformities into four types of imaging differences and it is widely accepted.

Several operative techniques to release the synostosis were published. However, a standard releasing technique and the most suitable time to release synostosis are still under research and evaluation.

In view of our case is within a toddler age group, she has a high probability of compensation by the shoulder and wrist joint function for her elbow deformities. Therefore, a thorough and multidisciplinary assessment is needed before commencing the best treatment option and subsequently proposed a holistic management protocol.

### CONCLUSION:

Congenital radio- ulnar synostosis is a rare malformation of the limbs. Management of synostosis is challenging and should be tailored to individual basis. The goal of management is to meet the minimal daily activity requirement of the patient. A multidisciplinary team involvement is advocated to minimize the consequences of this deformity.

### REFERENCES:

1. Lescault E, Mulligan J, Williams G: Congenital radioulnar synostosis in an active duty soldier: case report and literature review. *Mil Med*, 2000; 165(5): 425–28
2. Cleary JE, Omer GE., Jr Congenital proximal