

## RADIOULNAR SYNOSTOSIS IN A FATHER AND HIS 5 YEAR OLD DAUGHTER

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### ABSTRACT

Radioulnar synostosis is rare and its hereditary link is doubtful. In this report a 39-year old man with proximal bilateral radioulnar synostosis and his daughter with proximal unilateral radioulnar synostosis are presented.

**KEY WORDS:** Radioulnar synostosis.

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### INTRODUCTION

Sandifort originally described congenital radioulnar synostosis in 1793.<sup>1</sup> Congenital proximal radioulnar synostosis is a malformation caused by failure of normal prenatal separation of the radius and ulna. The persistent connection between the two bones is nearly always proximal; while distal radioulnar synostosis is extremely rare.<sup>2-6</sup> The connection is initially cartilaginous, and is not diagnosed until it ossifies, forming a bony synostosis. The forearm is usually fixed in pronation, probably because this is the normal fetal position. Functional deficits with congenital radioulnar synostosis depends on the severity of the deformity and on whether or not it is bilateral. In cases with severe fixed forearm pronation deformity, the patient is unable to compensate with scapular and glenohumeral motion. The forearm usually lies in the pronated or hyperpronated position.

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### CASE REPORT

A 5-year-old daughter with right proximal radioulnar synostosis visited our clinic. She was the second child of the family. The older child did not suffer radioulnar synostosis. Her 39 year old father had suffered proximal bilateral radioulnar synostosis. Thirty years ago, when he was nine year old he visited a hospital because of limitations in supinations and fixed pronations of the two radioulnar joint. After physical and radiological examinations bilateral radioulnar synostosis was diagnosed. Surgery was recommended immediately which could not be performed due to some problems. In the physical and laboratory examination, no other anomaly was found. He actively works as a vehicle driver. He has no important problem at work. However he has fixed arm pronation and limitation in supination in both sides (Fig I-III). His daughter showed proximal right radioulnar synostosis. Laboratory and physical examinations showed no evidence of other anomaly. On physical examination, there were a fixed pronation and limited supination. Flexion and extension movements were normal (Figure IV-VI).

### DISCUSSION

Congenital radioulnar synostosis is a rare condition with approximately 350 cases reported world wide.<sup>1</sup> There is three-to-two male predominance. Some researches reported



Figure-I: Fixed arm pronation in a 39 year old vehicle driver.



Figure-IV: Mild abnormality in the supination of the right forearm in 5 year old girl



Figure-II: Supination was limited in the both side in a 39 year old vehicle driver



Figure-V: Mild deformity in pronation of the right forearm in a 5 year old girl.



Figure-III: Proximal radioulnar synostosis in a 39 year old father as seen the plain x-ray.



Figure-VI: Proximal radioulnar synostosis as seen in the plain x-ray.

equal distribution in both and 60% of cases are bilateral.<sup>1</sup> Positive family histories have been reported.<sup>7-9</sup> Miura E et al.<sup>10</sup> reported 9% familial recurrence for the radioulnar synostosis. Although radioulnar synostosis is usually an isolated event, it may be associated with anomalies that affected renal, cardiovascular, central nervous system, and musculoskeletal systems. Renal anomalies involve anatomic malformations that can be screened by ultrasonography. Musculoskeletal problems include club feet, dislocated hips, polydactyly, syndactyly, and Madelung deformity.<sup>4-6,11</sup> Our patient did not show any other anomaly. Surgical intervention results remains controversial and frequently unsuccessful.<sup>1,12</sup> The first patient (father) is currently working as an active car driver. Due to adjustment to his condition, conservative treatment and exercises were recommended for him. Due to high failure rate of surgery and mild anomaly in the right hand and normal left hand in daughter, she was also recommended conservative treatment with follow up.

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