

## Three cases of congenital radioulnar synostosis followed for at least 29 years.

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### Abstract

Three patients with congenital proximal radioulnar synostosis who were followed for at least 29 years are presented. One patient underwent osteotomy to improve activities of daily living. At the time of the final follow-up, the carrying angle was improved. The other two

patients underwent radial head partial resection to prevent the locking due to synovial fold. Although lateral instability of the elbow was confirmed, osteoarthritis was not evident.

**Key words :**

### Introduction

Patients with a congenital proximal radioulnar synostosis (PRUS) have a bony bridge between the radius and the ulna, preventing forearm rotation. The forearms are fixed in an average of 30° to 40° of pronation. The connection is initially cartilaginous, and it eventually ossifies, forming a bony synostosis. The forearms are fixed in an average of 30° to 40° of pronation. Children with PRUS usually present between 3 and 6 years of age with painless limitation of forearm rotation and a slight flexion contracture.<sup>1</sup> PRUS usually does not cause significant functional impairment.<sup>2</sup> However, fixed pronation between 15° and 60° a relative indica-

tion, and forearm fixation in greater than 60° pronation a definite indication for surgery, if functional limitations are significant.<sup>3</sup>

We present herein three patients with congenital PRUS, without notable predispositions, who were followed for at least 29 years.

### Case report

#### Case 1

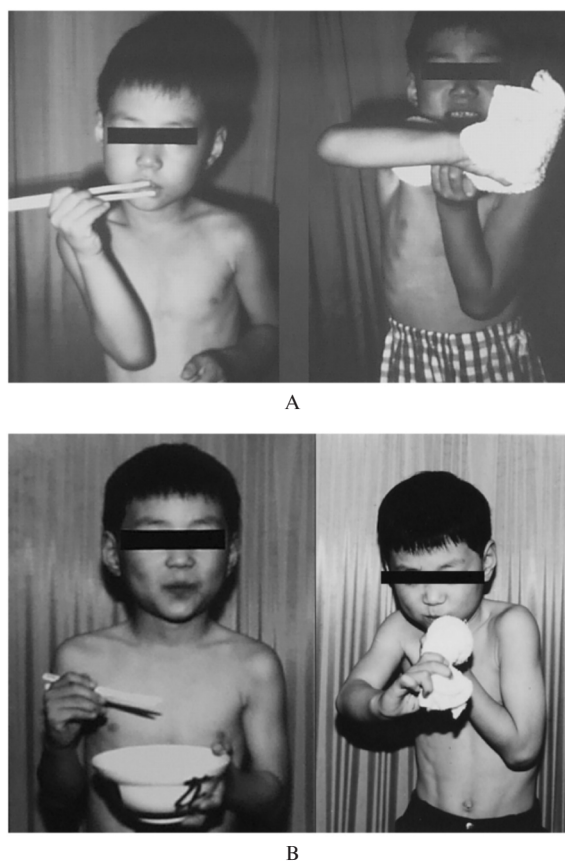
A 3-year-old child was consulted to our hospital. The boy complained of bilateral impaired forearm rotation and bilateral mobility limitation around the elbow at birth. Family and past medical histories were negative. X-ray showed PRUS with Wilkie I synostosis, in which the

medullary canals of the radius and ulna are



**Fig. 1** Case 1. A: X-ray of elbow joint at 3 year-old. B: X-ray of elbow joint at 7 year-old. C: An osteotomy was performed 1 cm proximal to the distal end of the synostosis to correct from 20° pronation to 20° supination. D: X-ray of elbow joint at 36 year-old.

joined, the proximal end of the radius is malformed and is fused to the ulna for several centimeters<sup>4</sup> (Fig. 1A). The patient was only followed up, as ADL were not impaired by employing compensatory movements of the shoulders and hands. At 7 years old, X-ray revealed no changes (Fig. 1B). However, the patient complained of difficulties wringing out towels and holding bowls (Fig. 2A). Both forearms were fixed in 20° pronation, and the range of motion (ROM) was 5-140° for the right and 10-145° for the left elbows, respectively. He was admitted to our hospital for surgery. In August 1980, the osteotomy, according to Green procedure was performed for the left forearm.<sup>6</sup> Brachial muscle was detached from radial bone. Osteotomy was performed 1 cm proximal to the distal end of the synostosis to correct from 20° pronation to 20° supination (Fig. 1C). Postoperatively, the patient improved ability to hold bowls and wringing out towels (Fig. 2B). At 20 years old, the



**Fig. 2** Case 1. A: Before surgery, both forearms were fixed in 20° pronation and holding bowls and wringing out towels are difficult. B: After surgery, the patient improved ability to hold bowls and wringing out towels.

**Table** Summary of 3 cases

	Age (Yrs,1)	Sex	Side 2)	Type 3)	Surgery (Yrs)	Modus	DASH score, 4)	Follow-up (Yrs)
							Final follow-up	
1	3	M	Blt	I	7	Osteotomy (Lt)	10.0	33
2	4	M	Blt	II(Rt) I(Lt)	16	Release (Rt)	11.4	40
3	6	F	Rt	II	8	Release (Rt)	6.8	29

1) Age at the first visit, 2) Affected side, Blt; bilateral, 3) Types of congenital radioulnar synostosis was evaluated on X-ray with Wilkie criteria, i.e. I: the medullary canals of the radius and ulna are joined, the radius is malformed and is fused to the ulna for several centimeters. II: the radius is fairly normal, but its proximal end is dislocated either anteriorly or posteriorly and is fused to the proximal ulnar shaft; the fusion is neither as extensive nor as intimate as in the first type. 4) DASH score: Disabilities of Arm, Shoulder, and Hand questionnaire (Ref. 5).

patient complained of a reduced quality of life due to an inability to play tennis or guitar, but ADL were maintained without additional surgery. His final visit was at 36 year-old. We evaluated shoulder and hand function and associated pain using the Disabilities of Arm, Shoulder, and Hand questionnaire (DASH); a DASH score of 0 corresponds to optimal function and a score of 100 represents maximal disability.<sup>5</sup> His DASH score was 10.0 points and X-ray showed the improved carrying angle from 170° to 150° (Table). The ulna had atrophied distal to the elbow, but osteoarthritis (OA) at the elbow was absent (Fig. 1D).

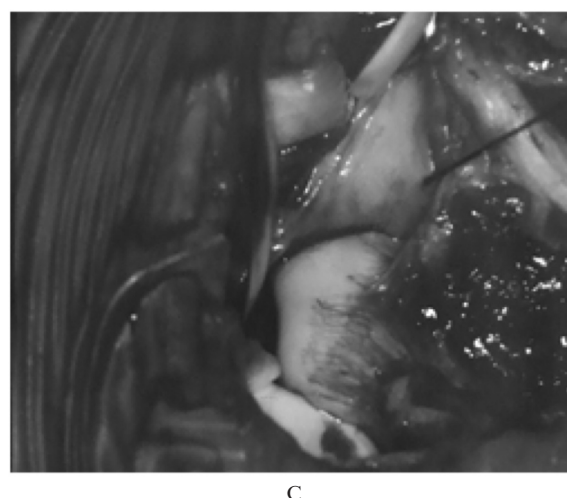
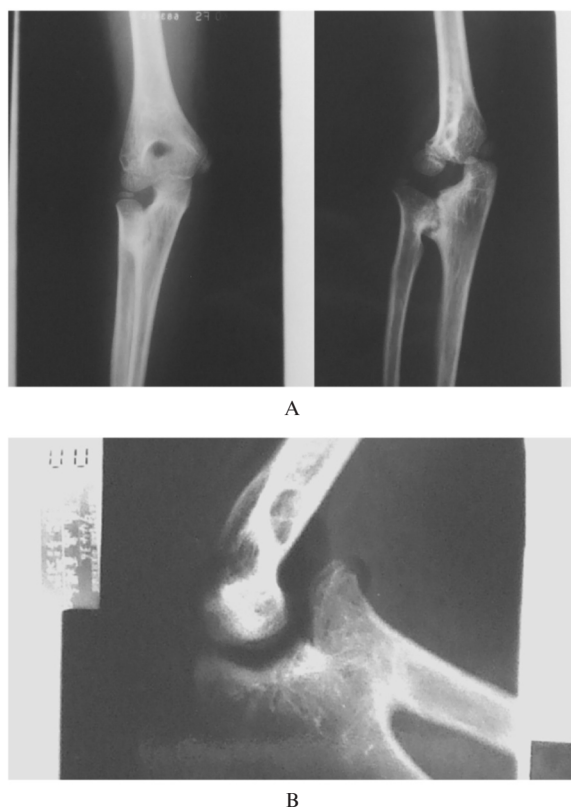
#### Case 2

A 4-year-old child was first consulted to our hospital. Bilateral congenital PRUS was confirmed with right Wilkie II synostosis, in which the radius is fairly normal, but its proximal end is dislocated either anteriorly or posteriorly and is fused to the proximal ulnar shaft; the fusion is neither as extensive nor as intimate as in the first type, with a 166° of carrying angle (Fig. 3A). On the other hand, X-ray showed Wilkie I synostosis with a 166° carrying angle. This patient was left untreated as ADL were not impaired. At 16 years old, the patient was visit again to our hospital and complained the right snapping elbow without any causes at the end of maximum right elbow flexion. Both forearms were fixed in the neutral position between pronation and supination, and ROM was 0-122° for the right and 0-146° for the left elbows, respectively. The locking phenomenon could be induced by forced right elbow flexion, and released by pas-

sive forced extension of the elbow with a clicking sound. Arthrography showed the anteriorly dislocated radial head compressed by a thickened synovial fold (annular ligament) (Fig. 3B). In July 1981, the right radial head was partially resected 7mm proximal to the distal end of the synostosis and release was carried out. During surgery, the synovial fold was confirmed to impinge on the radial head (Fig. 3C). His final visit was at 44 year-old. His DASH score was 11.4 points and X-ray showed that the forearm pronated 15° with 15° lateral instability, but the carrying angle showed no marked change, at 167°. ROM was 0-140°, and OA changes and recurrent snapping elbow were absent. Now, the patient complains of exertion elbow instability and upper extremity fatigue, but this can be addressed using a brace supporter (Table).

#### Case 3

A 6-year-old child was consulted to our hospital. The girl complained the swollen right dominant elbow joint without trauma. At 3 years old, the patient was unable to hold a spoon correctly, but no action was taken. X-ray showed the PRUS with Wilkie II synostosis (Fig. 4A). The carrying angle was 168°. However, the patient did not show any impairment of ADL and was thus followed up. At 8 years old, the patient displayed right snapping elbow after exercising, and underwent surgery because of the increased frequency of this event (Fig. 4B). The forearm was in the neutral position, and the elbow was in 18° valgus with ROM of 15-140°. As in Case 2, arthrography showed the dislocated radial head impinged by a thickened synovial



**Fig. 3** Case 2. A : X-ray of right elbow joint at 4 year-old. B : Arthrogram of right elbow joint at 16 year-old. C : During surgery, the synovial fold was confirmed to impinge on the radial head (Arrow). Proper release of ankylosis was done.

fold (Fig. 4C). In August 1982, partial radial head resection with release was performed for snapping elbow (Fig. 4D). At 15 years old, the right forearm showed 10° pronation, lateral instability was 20°, ROM was 15-140°, and the carrying angle was 158°. The patient played on a Softball team, but besides elbow pain after over-use, she did not experience any problems (Fig. 4E). At 41 years old, after 27 years surgery, radiography showed 30° forearm pronation and 15° lateral instability, but there were no marked changes in the carrying angle, which was 152°. ROM was 15-140°, and neither OA change nor recurrent snapping elbow was present. Although exertion elbow instability and upper extremity fatigue persisted, ADL were not impaired. Her DASH score was 6.8 points (Table).

### Discussion

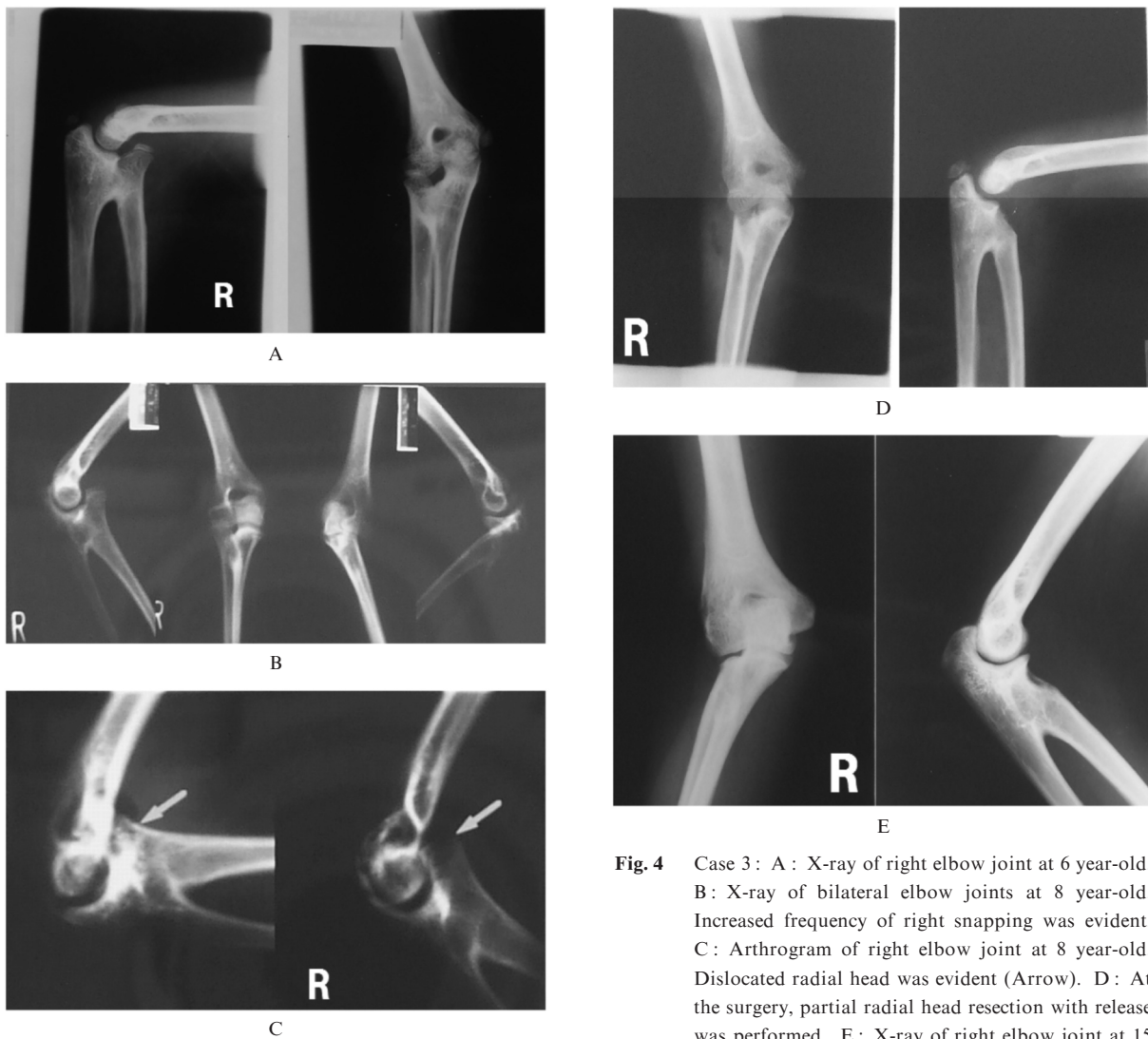
Most patients of congenital PRUS are not disabled enough to justify an extensive operation, because the impaired elbow movements can be compensated for by other joints. However, bilateral severely fixed pronation is accompanied with significant ADL limitation, as case 1. As well as ADL limitation, repeated snapping

between the dislocated radial head and thickened synovial fold was also the surgical indication.<sup>7</sup> Surgery for this condition is difficult. The fascial tissues are short and their fibers are abnormally directed, the interosseous membrane is narrow, and the supinator muscles may be abnormal or absent. Two major surgical procedures are commonly used in recent years. One is a derotational osteotomy to correct the forearm position.<sup>8,9</sup> In Case 1, osteotomy changed the forearm position from pronation to supination and increased stress on the radius. As a result, distal to the elbow, the radius mostly developed while the transverse diameter of the ulna decreased.

The other major surgical procedure is the surgical release.<sup>7</sup> All ADL tasks can also reportedly be carried out by compensatory movements in patients for whom the elbow is fixed in a near neutral position.<sup>10</sup> Kanaya et al. introduced a method of surgical release for congenital PRUS to improve the range of rotational movement, but this method is highly technically demanded and only short term follow up, i.e. less than 4 years, was reported.<sup>11</sup>

We obtained excellent result and no recurrence of synostosis with release and partial resection in





**Fig. 4** Case 3 : A : X-ray of right elbow joint at 6 year-old. B : X-ray of bilateral elbow joints at 8 year-old. Increased frequency of right snapping was evident. C : Arthrogram of right elbow joint at 8 year-old. Dislocated radial head was evident (Arrow). D : At the surgery, partial radial head resection with release was performed. E : X-ray of right elbow joint at 15 year-old.

case 2 and 3. However, marked instability was noted. Instability could have been minimized by minimizing radial head resection ; resecting only the synovial fold and elongating the annular ligament.<sup>12</sup> As the elbow is not a weight-bearing joint, degenerative changes were not seen, but follow-up will be necessary.

To our best knowledge, this report is the longest follow-up of PRUS. In these three patients surgical objectives were achieved and no complications, such as OA, were encountered.

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