Skeletal Radiol (2009) 38:585–591
DOI 10.1007/s00256-008-0638-x

SCIENTIFIC ARTICLE

The carpal bones in Poland syndrome

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Received: 28 October 2008 / Revised: 17 December 2008 / Accepted: 19 December 2008 / Published online: 30 January 2009

Abstract
Objective Classical Poland syndrome is represented by unilateral aplasia of the sternocostal head of the pectoralis major muscle and ipsilateral simple syndactyly and brachydactyly. Various classifications of the severity of hand involvement have been proposed. Since its initial description, numerous studies have been made of the bony, soft tissue, organ, and hematological disturbances. However, carpal bone involvement has been largely overlooked. The purpose of this study was to evaluate the carpal bones in patients with Poland syndrome from a local (Manitoba) cohort as well as those from the literature.
Materials and methods Hand radiographs from local patients and cases identified from the literature with confirmed Poland syndrome were examined for evidence of carpal bone involvement. Only cases with radiographs of adequate quality were included in the analysis. Clinical information (including gender and age) was necessary for evaluation of bone maturation. In total, seven local patients and 23 patients from the literature were evaluated. Ethics approval for study of the local patients was obtained by the Research Ethics Board of the University of Manitoba.
Results Of the 23 literature patients, 12 patients (52%) had abnormal findings. Of the abnormal patients, four of 12 (33%) had carpal fusions, eight of 12 (67%) showed disharmonious ossification between the carpal and tubular bones and seven of 12 (58%) showed delay of carpal ossification. Of the local cohort, three patients were too young to characterize carpal involvement. Of the four remaining patients, two (50%) had abnormal carpal morphology, three out of four had disharmonious ossification and all four had delay of ossification of carpal bones. Carpal fusions, particularly of the scaphoid and trapezium, were common in both groups.
Conclusion Carpal bone anomalies (delay, disharmony, and/or fusions) are frequent in Poland syndrome and can occur in patients with either mild or severe hand involvement. Imaging of the unaffected hand is helpful in determining the extent of carpal findings.

Keywords Poland syndrome · Carpal fusion · Carpal coalition · Scaphoid · Trapezium

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Introduction

Classical Poland syndrome is represented by unilateral aplasia of the sternocostal head of the pectoralis major muscle and ipsilateral simple syndactyly and brachydactyly. Since its description by Poland in 1841 [1], numerous studies have been made of the various bony, soft tissue, organ, and hematological disturbances [2]. The carpal bones have been largely overlooked. In our review, brief references [3, 4] and one case study [5] were identified that specifically addressed carpal bone involvement in Poland syndrome. The purpose of this study was to evaluate the carpal bones in patients with Poland syndrome.

Materials and methods

All patients with Poland syndrome whose involved hand had been radiographed at the Health Sciences Center were reviewed. Ethics approval for study of the local patients was obtained by the Research Ethics Board of the University of Manitoba. A search of the literature using Pubmed with the term "Poland Syndrome" was performed and relevant papers were examined for hand X-rays and documentation of carpal bone anomalies. All published X-rays of adequate quality were examined for evidence of carpal bone abnormalities. In addition, six textbooks were utilized [6–11]. In order to assess bone maturation, patient gender and age were necessary clinical information.

Results

Local cases

A total of seven local patients had hand radiographs available for examination (five males, two females). Three of the seven showed no obvious abnormalities; however, these patients were only 5 days, 1 month, and 7 months of age, respectively. Anomalies may become evident later in development. The findings from the other four patients are summarized in Table 1. Delayed ossification was evident in all four patients. Patients 1, 3, and 4 showed delay of both carpal and tubular bones of the hands. Patient 1 had symmetrical ossification delay of tubular and carpal bones (Fig. 1a,b). Patient 1 had ipsilateral radioulnar synostosis proximally (Fig. 1c). Patients 2, 3, and 4 showed disharmonious ossification between carpal and tubular bones (Figs. 2a,b and 3). Patient 2 had ipsilateral posterior dislocation of the radial head. Two patients had carpal fusions (Figs. 1a,b and 2b). For Patient 2, there were initially separate ossification centers for the scaphoid and trapezium (Fig. 2a) that later fused (Fig. 2b).

Literature cases

Twenty-three patient radiographs from the literature were of sufficient quality to assess carpal bone involvement [5–20]. Of the 23 literature patients, eleven (48%) had completely normal-appearing carpal bones [7–9, 12, 14] and twelve had abnormal findings [5, 6, 10, 11, 13, 15–20]. Of the abnormal patients, four had fusions [5, 10, 11, 19], eight had disharmonious ossification between carpal and tubular bones [6, 13, 15–18, 20] and seven had delay of carpal bone ossification [6, 13, 15–18, 20] (Table 2). An additional 24 radiographs from a paper by König and Lenz [4] are discussed separately due to the fact that accompanying clinical information (gender and chronological age) was not available.

König and Lenz cases

Figures 2, 3, 4, 5, 10, 12 and 13 from the paper by König and Lenz [4] revealed normal carpal bones. Figures 1, 7, 8, 9, 11, 15, 16, 22, 24 were of insufficient quality for assessment.

Table 1 Local Poland patients with abnormalities

Patient number	Patient's age	Patient's gender	Affected side	Carpal anomaly
1	9 years 6 months	F	Left	Scaphoid—trapezium fusion Lunate—triquetrium fusion Hypoplastic trapezoid Bone age 6 years 10 months–7 years 10 months in all bones[a] Decreased carpal height relative to unaffected hand Proximal radioulnar fusion (Fig. 1a,b,c)
2	12 years	M	Left	Scaphoid–trapezium fusion Hypoplastic trapezoid Carpal bone age 9 years–10 years Tubular bone age 12 years[b] Abnormally small inter-carpal spaces Posterior dislocation of the radial head Fig. 2a,b
3	4 years 6 months	M	Right	Carpal bone age 2 years Tubular bone age 3 years (Fig. 3)
4	3 years 6 months	M	Right	Carpal bone age 1 year 6 months Tubular bone age 2 years–2 years 8 months

[a] At 4 years 6 months, the carpal bones were 1 year 6 months–2 years and the tubular bones were 2 years 6 months
[b] At 4 years of age this patient's carpal bone age was 1 year–1 year 9 months and the tubular bones were 2 years 8 months. At 7 years, the bone age was 4 years 6 months for both carpal and tubular bones

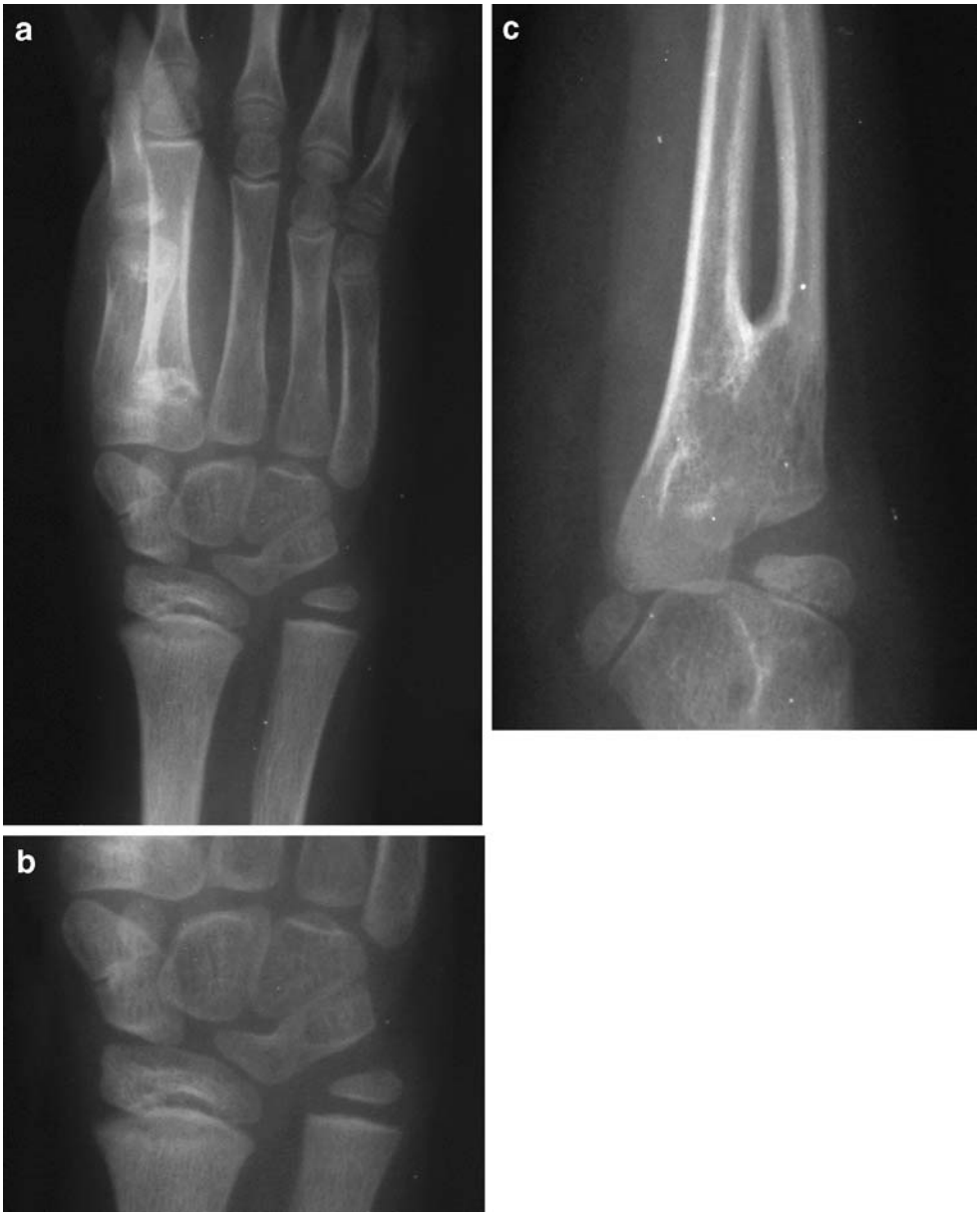

Fig. 1 **a**,**b** Patient 1. Female patient at 9 years 6 months with affected left upper extremity. There is decreased bone age of all bones. There is a pseudoepiphysis at the base of the second metacarpal. The carpal height is decreased and there is scaphoid–trapezium fusion, lunate–triquetrum fusion and a hypoplastic trapezoid. There is an ulnar plus deformity. **c** This patient also had radioulnar synostosis proximally

The seven radiographs that were of adequate quality for assessment and had anomalies are summarized in Table 3. Four patients had carpal fusions.

Discussion

Classical hand findings

Poland syndrome is hypothesized to originate from a vascular insult, specifically of the subclavian artery, at 6 weeks gestation [21]. The classical findings in the hands of Poland syndrome patients are simple syndactyly and brachydactyly. Gausewitz [22] divided the hand anomalies into four subcategories: type 1—presence of all five digits, (that may be hypoplastic); type 2—the central digits are absent while the border digits are present; type 3—no functional digits; type 4—radial ray defects with an absent thumb. More recently, Al-Qattan proposed a more detailed classification, comprising seven different types, taking into account a wider range from a normal hand to a phocomelia-like deficiency [14]. Soft tissue hand anomalies have also been described including absence of flexor and extensor tendons and muscle anomalies [9].

Carpal findings

Carpal anomalies observed in our study were: disharmonious ossification between carpal and tubular bones, carpal bone maturation delay, and carpal coalition. Carpal fusions were

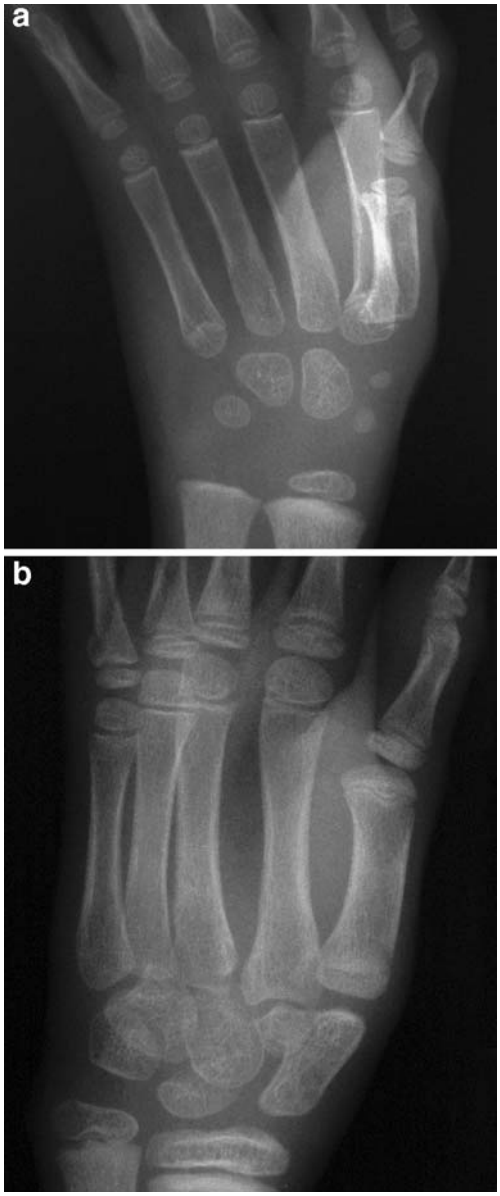

Fig. 2 **a**. Patient 2. Male patient with the left hand affected. Patient at 7 years of age. There is a pseudoepiphysis at the distal end of the first metacarpal and one at the base of the fifth metacarpal. The carpal height is decreased. There are separate ossification centers of the scaphoid and trapezium, that later fused as evidenced by **b**, which was taken at 12 years 6 months. **b** The pseudoeipiphysis at the distal end of the first metacarpal remains but the one at the base of the fifth metacarpal is now fused. There is an ulnar plus deformity

present in two local cases and four literature cases. In all these cases, the fusion involved the scaphoid and the trapezium. Some patients had multiple fusions.

Findings from König and Lenz

König and Lenz reviewed 66 cases of Poland syndrome and included 56 in their final analysis [4]. Their classification ranged from the mildest form—brachydactyly/

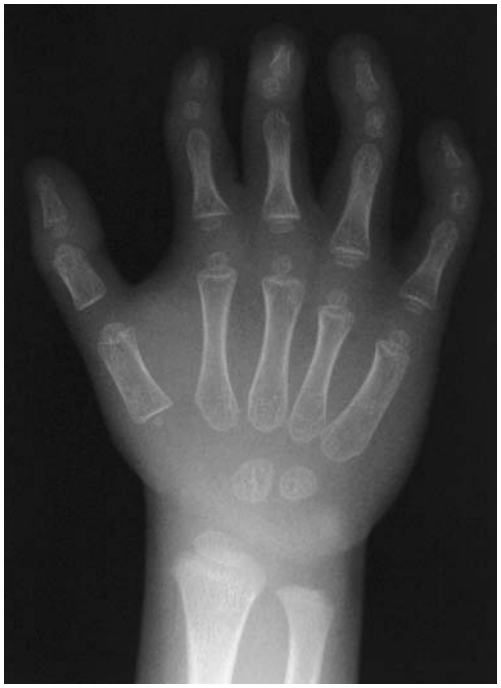

Fig. 3 Patient 3. Male patient at 4 years 6 months. Right hand. Disharmonious ossification. Carpal bone age 2 years, tubular bone age 3 years. The middle phalanges are shortened and there is a pseudoepiphysis at the distal end of the first metacarpal

symbrachydactyly (33 cases), to atypical split hand (28 cases), to the most severe form—peromelia (5 cases). For the symbrachydactyly patients, only phalanges were affected; in the atypical split hand group, metacarpals were affected, and carpal bones could be absent in the most severe form. However, from the 24 published radiographs, carpal involvement (e.g., coalition) was present in some of the milder cases. Scaphoid–trapezium fusions were present in three cases. Age and gender were not provided for any of the radiographs, therefore, bone maturation has not been commented on.

Gender and side predilection

Of the 23 literature patients, the gender was reported for 19. Fourteen were male (74%) and five were female (26%). Of the local cases, five of the seven (71%) were male. Sporadic Poland syndrome is more common in males. The reason for this discrepancy is unknown [22, 24]. Poland syndrome shows a predilection for the right side in males, there is no preference for either side in females [23, 25]. This may be since the development of the female embryo lags behind that of the male during the period in which laterality is defined [23, 26]. Inherited cases show no preference for either side or gender [23, 27]. It has been postulated that the factors involved in inherited Poland syndrome act at an earlier stage, prior to lateralization, than those involved in

Table 2 Literature patients with carpal anomalies

Patient's age (years)	Patient's gender	Affected side	Source	Carpal anomaly
2[a]	M	Left	Al-Qattan [15] Fig. 1b	Carpal bone age 3 months Tubular bone age 2 years
5	M	Right	Atar [17] Case2	Carpal bones approximately 2 years Tubular bones approximately 5 years
2	F	Right	Foucras [18]	Carpal bone age between 3–6 months Tubular bone age 1 year–1 year 3 months (phalanges <1 year)
13	M	Left	Guion-Almeida [19]	Scaphoid–trapezium fusion Absent hamate Absent trapezoid Hypoplastic lunate
26	M	Left	Hadely [5]	Scaphoid–trapezium fusion Lunate–triquetral fusion
?	F	Right	Karev [16] Fig. 15	Tubular bone age 2 yrs 6/12 Hypoplastic capitate and hamate (for 2 yrs 6/12), narrow carpus
5	M	Right	Gordon [20]	Carpal bone age 2 years bilaterally Tubular bone age 3 years–3 years 5 months bilaterally.
2 years 6/12	M	Left	Kabra [13]	Carpal bone age 9 months–1 year Tubular bone age 1 year 6/12
Adult	M	Right	Gupta [10]	Scaphoid–trapezium fusion
3 years 10/12	M	Right (assumed)	Poznanski [6] Fig. 13–292	Tubular bones 4yrs–4yrs 6/12 Carpal bones 3 years 6/12 Missing triquetrum, hypoplastic lunate
?	?	Right (assumed)	Poznanski [6] Fig. 13–294	Tubular bone age 1 yr 3/12–6/12 Carpal bone age 2 years 6/12–8/12
Adult	M	Left (assumed)	Stevenson and Hall [11] Fig. 20–21	First metacarpal/scaphoid/trapezium fusion, hamate/triquetrum fusion, capitate/hamate fusion

[a] Chronological age was not given, overall bone age was used as an estimate.

Table 3 Summary of carpal findings from published radiographs of König and Lenz

Figure	Carpal findings
6	Capitate–hamate fusion Hypoplastic trapezoid
14	Scaphoid–trapezium fusion Trapezoid–capitate fusion Abnormally rounded lunate
17	Absent trapezoid Hypoplastic trapezium
18	Scaphoid–trapezium fusion Hamate–triquetrum fusion Absent trapezoid Absent capitate
19	Scaphoid–trapezium fusion Very hypoplastic lunate
20	Absence of all carpal bones except capitate and hamate (young patient)
21	Two carpal bones present—likely hamate and very hypoplastic trapezium

sporadic cases [23]. Of the literature patients with carpal anomalies, there was no side predilection for involvement (7/12 right side, 5/12 left side involvement). This was similar for our local cases with carpal involvement (two—left side, two—right side).

Hypoplasia

Ireland et al. noted that 36 out of a series of 43 patients with Poland syndrome had carpal hypoplasia [3]. Imamura et al. noted that 13 of their patients with symbrachydactyly, nine of whom were believed to have Poland syndrome, had carpal hypoplasia [28]. Ireland et al. did not report their criteria for carpal hypoplasia. Imamura et al. used the patients' contralateral hand for comparison. In one local case (Patient 1), a radiograph of the unaffected hand was available. The carpal bones of the affected hand were hypoplastic relative to the unaffected hand. As radiographs of the unaffected hand were largely unavailable for our study and no satisfactory method of measuring carpal size could be applied, carpal size has not been commented on in the other patients.

Disharmonious bone maturation and delay

In one local case (Patient 1), there was uniform delay of all hand bones. In three of the seven local patients (Patients 2, 3, and 4), the carpal bones were more delayed than the tubular bones. In seven of the published cases, the carpal bones were more delayed than the tubular bones [6, 13, 15–18, 20], in three cases, there was delay of both carpal and tubular bones [13, 18, 20]. Disharmonious maturation of the hand has been described in various congenital malformation syndromes, metabolic disorders and skeletal dysplasias [29]. Assigning bone ages in such situations can be problematic. However, the authors note that the degree and pattern of disharmony may be of diagnostic benefit. Imamura et al. found no cases of delay in their cases of symbrachydactyly [28]; however, all the patients in their study were over 10 years old, with an average age of sixteen. The patients in our study who had delay were all younger than ten, except for Patient 2, who was twelve. There were two local patients, Patients 1 and 2, for whom hand X-rays at more than one age were available. In patient 1, the carpal and tubular bone ages were delayed at both 4 years 6 months and 9 years 6 months of age. Patient 2 had hand X-rays at 4, 7, and 12 years of age. By 12 years, the tubular bones (not carpal bones) had caught up to the patient's chronological age. In his paper on bone ages of congenitally anomalous hands, Araki concluded that delay should improve with age [30]. The findings in Patient 2 support Araki's conclusion.

Carpal fusion

Carpal coalition has been proposed to result from incomplete separation of cartilaginous precursors during the embryonic period [31]. Figures 2a and 2b demonstrate that initially, there were separate ossification centers of the scaphoid and trapezium, which later fused. In the general population, carpal coalition occurs in approximately 3.6% of people [32]. Carpal fusion has been found to be more common in those of West African descent, at a rate of approximately 8%, with lunate–triquetral being the most common [33]. Garn et al. found that in four out of five cases of carpal coalition, only a single pair of carpal bones was involved [32]. Of the four literature cases with carpal fusion, two had more than one pair of carpal bones involved, as did one local case. Of the four patients with carpal fusion from the König and Lenz paper, two patients had two fusions each. According to Thijn, carpal fusion that crosses the carpal rows is usually associated with a congenital syndrome whereas fusion between carpal bones in the same row is usually an isolated anomaly [34]. All cases of fusions examined in this paper cross rows supporting Thijn's theory. The results of our study suggest that carpal coalition, usually of the scaphoid and trapezium, is more common in Poland syndrome than in the general population.

Spectrum

The hand anomalies in Poland syndrome are not limited to simple syndactyly and brachydactyly, but rather occur on a spectrum. This includes virtually normal to atypical cleft hands, to ray defects and even peromelia [4, 14, 22]. It is therefore not surprising that carpal anomalies can occur in Poland syndrome. Interestingly, the same anomalies that occur in the more severely affected patients may also be seen in milder cases.

Conclusion

Delayed carpal bone maturation, disharmonious ossification and carpal coalition, particularly of the scaphoid and trapezium, are characteristic of Poland syndrome and have not previously been fully appreciated. Imaging of the unaffected hand can be helpful in determining the extent of carpal involvement.

Acknowledgement The authors wish to thank Dr. Mark Torchia for valuable discussion.

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