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Postaxial polydactyly of the hand in Japanese patients: Case series reports

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Summary *Purpose:* The incidence of postaxial polydactyly of the hand is rare in Japan. This study aimed to compare the clinical presentation of postaxial polydactyly between a cohort of patients from Japan and those from other racial and ethnic backgrounds.
Methods: In this retrospective study, we included 30 patients who were treated at our hospital during a 25-year study period (1990–2015). Based on the clinical records of these patients, we characterized the Japanese presentation of the condition. We searched for studies that included other racial and ethnic groups and characterized the clinical presentations. Then, we compared the clinical presentations between Japanese patients and other racial and ethnic groups.
Results: A total of 19 male and 11 female patients were treated in our hospital, and bilateral and unilateral involvements (right side: 4 patients; left side: 4 patients) were observed in 22 and 8 patients, respectively. Moreover, 22 postaxial polydactylies were type A and 28 polydactylies were type B, which were classified using the Temtamy–McKusick classification system. In addition, 4 patients had a family history of hand postaxial polydactylies; 18, 6, 4, and 3 patients presented with polydactyly of the foot, syndactyly, systemic abnormalities, and related syndromes, respectively.
Conclusion: Japanese patients had two distinguishing characteristics: (1) when the condition was unilateral, left side and right side involvement was almost equal with regard to incidence and (2) associated polydactylies of the foot were more common (60%) than those in other

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cohorts (less than 31%). To better understand postaxial polydactyly of the hand, guidelines to record the clinical presentations in patients with such a condition must be developed.

Introduction

Polydactyly of the hand is one of the most common congenital abnormalities of the hand[1-3]. The clinical presentations of the disorder are categorized into three groups according to the position of the duplicated digit: preaxial (radial), central, or postaxial (ulnar). The position of the duplicated digit is more likely different among individuals of various racial or ethnic backgrounds. Moreover, postaxial polydactyly is significantly more common in patients of African descent[2], and preaxial polydactyly is most common in Caucasian patients[2]. Among Japanese patients, preaxial polydactyly is the most common type, which accounts for approximately 90% of all polydactylies of the hand[4,5]. Postaxial polydactyly is rare in individuals with Japanese ancestry, which accounts for 0–13.2% of all polydactylies of the hand[4,6,7]. Oka et al. reported in their study that the incidence of postaxial polydactyly of the hand among newborn children in the Miyagi prefecture (one of the prefectures in Japan) was 0.8/10,000[6]. Moreover, this result indicated that postaxial polydactyly of the hand is rare among Japanese individuals compared with those of African descent (1/100–1/300), Caucasian (1/1500–1/3300), and Middle Eastern (1/1000)[2,8,9]. Case series of postaxial polydactylies of the hand in Japanese are also limited, and most studies included a small number of patients[10,11].

The presentation of postaxial polydactyly differs according to patients' racial and ethnic backgrounds. A number of published reports have compared the clinical presentation of the disorder mainly between patients of African and Caucasian descent[1,9,12]. Recently, a case series describing postaxial polydactyly in patients of Middle Eastern descent was published[8]. On the basis of this report, the presentation of postaxial polydactyly in individuals of Middle Eastern descent was between that of individuals of African and Caucasian descent. Therefore, the presentation of postaxial polydactyly of the hand in Japanese patients may be different and unique from that of individuals of Caucasian, African, and Middle Eastern descent. This study was conducted to compare the clinical presentations of postaxial polydactyly in a cohort of Japanese patients with those of other racial and ethnic backgrounds. This study also investigated whether the cohort of Japanese patients had unique characteristics.

Patients and methods

All study-related procedures received complete institutional review board approval from the ethics committee of our hospital. For this retrospective review, we obtained the medical records of patients who were surgically treated for postaxial polydactyly of the hand at our hospital during the 25-year study period between 1990 and 2015. We included all patients who were treated at our hospital but excluded those whose medical records were incomplete. We recorded each patient's clinical and radiological data, including sex, unilateral (right or left) or bilateral involvement, family history (including any degree of consanguinity), associated hand or foot abnormalities, systemic abnormalities, related syndromes, and classification of polydactyly.

Table 1 Temtamy-McKusick classification.

Type A: the extra digit is well formed and has an articulation
Type B: the extra digit is poorly formed and is connected to the hand by a skin bridge

To compare the characteristics of patients from Japan with those of individuals of other racial and ethnic backgrounds, we searched for reports published worldwide after 1997. We used PubMed to search for reports published in English using the keywords "postaxial polydactyly" and "ulnar polydactyly." The inclusion criteria were as follows: case series of postaxial polydactyly of the hand (not including both hand and foot postaxial polydactyly), those including more than 30 patients, and those including comparison items, as shown below. From these case series[1,2,8,13,14], we established four racial and ethnic groups (Japanese, Caucasian, African, and Middle Eastern descent) to compare the clinical features of postaxial polydactylies of the hand. To classify postaxial polydactyly of the hand, we used the Temtamy-McKusick classification system (Table 1)[15]. In cases where other classification systems were used, we changed the data to conform to the Temtamy-McKusick classification system. The comparison items were as follows: sex, affected side, classification, family history, associated polydactylies or syndactylies of the hand and foot, associated systemic abnormalities, and associated syndromes.

Results

During the 25-year study period, 30 patients with 52 postaxial polydactylies were surgically treated in our institution (Table 2). The results of the studies conducted in Austria, USA, Turkey, and Saudi Arabia series are shown in Table 3[1,2.8,13,14]. The comparisons of each category among five studies are shown in Tables 4–9.

Discussion

Using the results, we compared the clinical forms of postaxial polydactyly of the hand among individuals of four different racial and ethnic backgrounds: Japanese, Caucasian,

Table 2 Results of our series data.

	Number of cases	Sex		Affected side			Classification[a]		Family history of ulnar polydactyly	Associated polydactylies of the hand	Associated polydactylies of the foot	Associated syndactylies of the hand and foot	Associated systemic abnormalities	Associated syndromes
		Male	Female	Right	Left	Bilateral	A	B						
Our series	30	19	11	4	4	22	22	28	4[b]	0	18	6	4[c]	3[d]

[a] In one patient: there were no records of classification.
[b] All four patients had bilateral postaxial polydactyly of the hand.
[c] Three cases with cardiovascular disease, and one case with genitourinary disease.
[d] Two cases with Ellis-van Creveld syndrome, and one case with trisomy13 and 18.

Table 3 Results of studies conducted in other countries.

	Number of cases	Sex		Affected side			Classification		Family history of ulnar polydactyly	Associated polydactylies of the hand	Associated polydactylies of the foot	Associated syndactylies of the hand and foot	Associated systemic abnormalities	Associated syndromes
		Male	Female	Right	Left	Bilateral	A	B						
Austria	32	20	12	2	10	20	12	41	0	1	9		1	6
Saudi Arabia	94	41	53	15	35	44	32	62	10	2	28	6	13	6
Turkey	42	22	20	10	12	20	23	39	8	3	14	4	9	1
USA	148	85	63	24%	76%		26	122	49	5	20	11	16	5
(African descent)	103			32		71	6	97						0

Blank cells: There were no data in literature.

Table 4 Comparison of "sex".

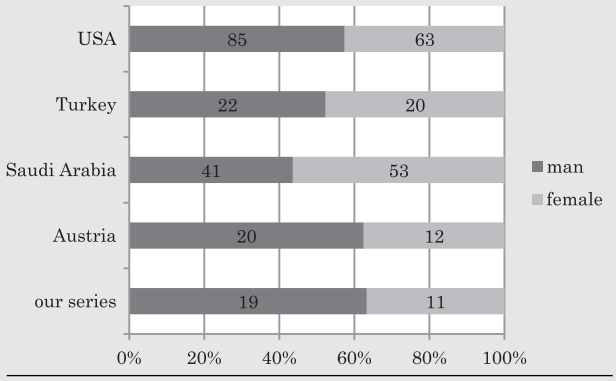
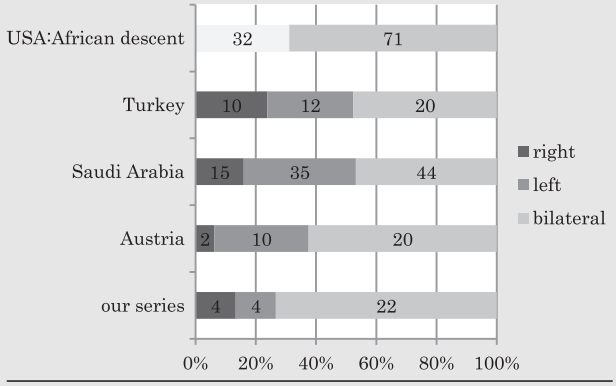


Table 5 Comparison of "affected side".



In the USA series, there were no data of unilateral vs. bilateral. Among unilateral cases, patients with right side involvement were 24% and those with left side involvement were 76%. For the African descent patients in the USA series, only unilateral vs. bilateral data were given, but there were no details about right side or left side involvement.

Table 6 Comparison of type of postaxial polydactyly of the hand (By Temtamy-McKusick classification).

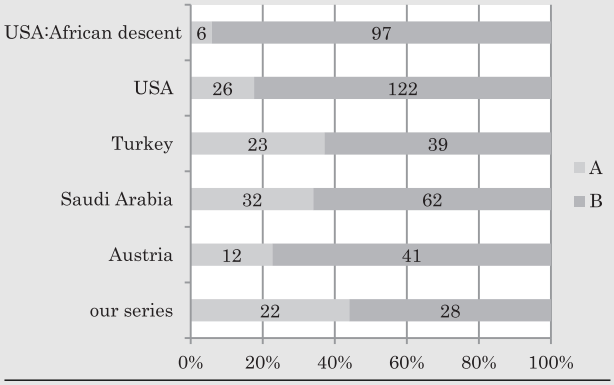
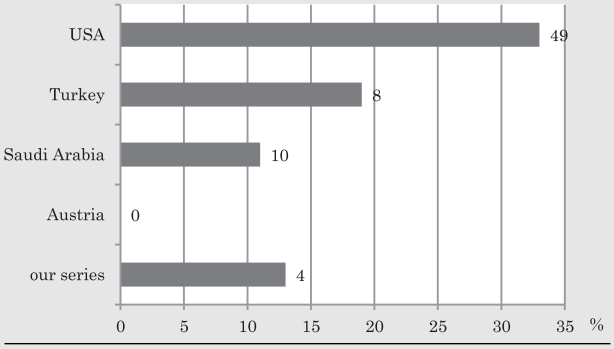


Table 7 Comparison of family history of ulnar polydactyly.



In the Austria series, 41% (13 patients) had a positive family history of other types of polydactyly. In the USA series, family histories of 87 patients were unknown. There might be more patients with a positive family history. All the patients with a positive family history in our series, Turkey series, and Saudi Arabia series were involved bilaterally.

Middle Eastern, and African descent. The results are summarized in Table 10. The data obtained from the study conducted in Austria were representative of Caucasians, and those obtained from studies conducted in Turkey and Saudi Arabia were representative of individuals of Middle Eastern descent. The data were presented as mean values of percentages for ease of understanding.

According to the results shown in Table 10, the unique clinical features of postaxial polydactyly of the hand in Japanese patients were as follows: (1) when the condition was unilateral, left side and right side involvements were equal with regard to incidence; and (2) associated polydactylies of the foot were more common than other clinical forms. These two characteristics almost coincided with those of prior reports on Japanese postaxial polydactyly of the hand[6,10,11].

Moreover, other characteristics of the presentation of postaxial polydactyly were revealed. In terms of presentation in individuals of Middle Eastern descent, the number of female patients was higher than that of male patients, and the number of individuals with unilateral involvement was higher than that of individuals with bilateral involvement. These two points were not observed in other clinical forms, which are supported by the report of Al-Qattan.[8] In terms of presentation in individuals of African descent, type B polydactyly was significantly more common than type A and cases wherein related syndromes were rare. The number of individuals of Japanese and Middle Eastern descent with a family history of postaxial polydactyly of the hand was almost equal.

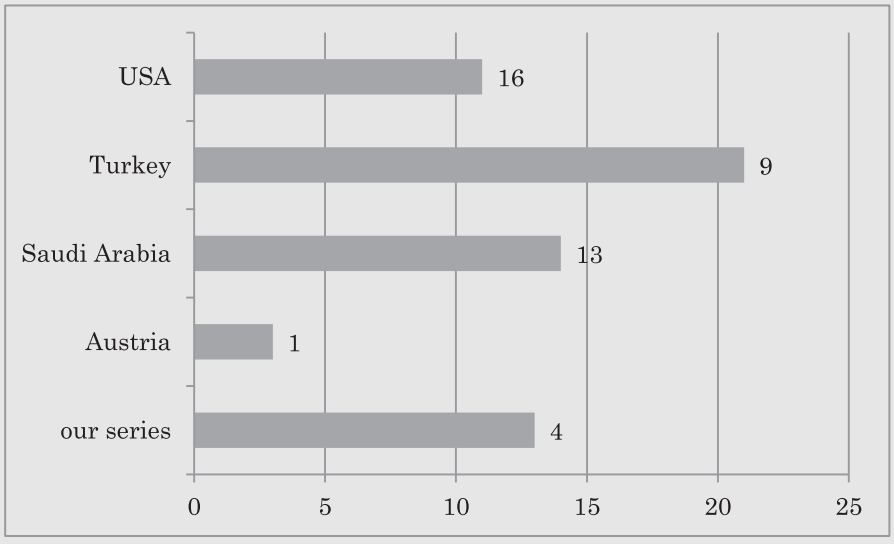
Although several reports on postaxial polydactyly are available, numerous previous studies have reported the differences between populations of Caucasian and African descent[12,16]. Moreover, the majority of these studies included not only postaxial polydactyly of the hand but also postaxial polydactyly of the foot[12,16-18]. In 1997, Castilla reported that postaxial polydactylies of the hand and that of foot have epidemiologically different congenital features[14]. Thereafter, studies that included only postaxial polydactyly of the hand have been published. Therefore,

Table 8 Comparison of associated syndromes.

	Our series	Austria	Saudi Arabia	Turkey	USA[a]
Ellis-van Creveld syndrome	2	2			
Trisomy 13, 18	1				
Trisomy 13				1	1(Caucasian)
Jeune syndrome		2			
Laurence-Moon-Biedl syndrome			3		
Greig syndrome			2		
Apert syndrome			1		
Carpenter syndrome					2(Caucasian)
Oral-facial syndrome					1(Caucasian)
Holt-Oram syndrome					1(Asian)
Unclassified dysmorphic syndrome		1			

[a] There were no African descent patients with any associated syndromes in the USA series.

Table 9 Associated systemic abnormalities.



associated systemic abnormalities	cardiac	gastrointestinal	central nervous system	renal	genitourinary	endocrine	ocular	extenal ear	others
our series	3				1				
Austria		1							
Saudi Arabia	4			5				2	2
Turkey	2		3				3		1
USA	1	1	4		3	1	2	2	2

Associated systemic abnormalities: musculoskeletal abnormalities were excluded.

we searched for reports published after 1997 worldwide and found four reports that included only postaxial polydactyly of the hand[1,2,8,13]. Al-Qattan reported the comparisons of presentation of postaxial polydactyly of the hand between individuals of Middle Eastern descent and those of African and Caucasian descent[8]. In their study, they established a clinical form for individuals of African and Caucasian descent by referring to other reports, with each parameter of the clinical feature described using the mean percentage of other reports. They statistically analyzed the three clinical features of individuals of Middle Eastern, Caucasian, and African descent; however, accuracy of this type of analysis

Table 10 Comparison of Japanese patients with patients of other racial and ethnic backgrounds.

	Number of cases	Sex		Affected side			Type		Family history ulnar polydactyly	Associated polydactylies of the hand	Associated polydactylies of the foot	Associated syndactylies	Associated systemic abnormalities	Associated syndromes
		Male	Female	Right	Left	Bilateral	A	B						
Japanese	30	63	37	13.5	13.5	73	44	56	13	0	60	20	13	10
Caucasian	32	63	37	6	31	63	23	77	0	3	28		3	19
Middle Eastern	136	46	54	18	35	47	35	65	13	4	31	7	16	5
African descent	103			31		69	6	94						0

Blank cells: There were no data in literature.
The data are presented as mean values of percentages.

was questioned. In all the reports that were assessed, no precise details about all the categories of the clinical features were obtained. This result was similar to that of our study, which made the accurate comparison of the different clinical presentations extremely challenging.

Because of several limitations of this study, we could not perform a precise statistical analysis. Therefore, our conclusions are probably not accurate. First, the four reports we studied for comparisons included fewer than 150 cases, and our series and the Austria series included only approximately 30 cases. All the reports (including that of our series) were studied in a single center. To make a precise clinical presentation of each ethnical group, more cases should be studied in a multicenter study. Second, some reports (particularly in the series of studies conducted in the USA[1]) included a heterogeneous mix of patients from various racial backgrounds. Third, in our facility, only patients who underwent surgeries for postaxial polydactylies were included. The number of patients with type B postaxial polydactyly whose fingers were ligated in maternity hospitals may be higher. Fourth, we included any degree of consanguinity to indicate a positive family history. However, this did not apply to all reports used for comparison, particularly because some reports did not mention the degree of consanguinity that had been included. Fifth, data of individuals of African descent included those of African-American descent in a few reports[1,9,12]. As this was extremely confusing, we referred to previous reports that included individuals of African or African-American descent. However, we could not precisely distinguish the data. Sixth, we considered the data obtained from the study conducted in Austria as a representative of individuals of Caucasian descent and those obtained from studies conducted in Saudi Arabia and Turkey as a representative of individuals of Middle Eastern descent on the basis of demographic characteristics. However, this consideration might not be accurate. Singer mentioned in their reports that the clinical presentations closely resembled those of individuals of Caucasian descent according to demographic characteristics[2]. We also considered the individuals of both the Saudi Arabia series and the Turkey series as representatives of the Middle Eastern descent for the same reason. However, this consideration might not be accurate. Thus, this remains a limitation of our study. Further studies must be conducted to validate this result.

Postaxial polydactyly of the hand is a disorder with different presentations in patients of different racial and ethnic backgrounds. Although it is interesting to compare the clinical features of patients of various racial and ethnic backgrounds, there are no guidelines on how to record clinical presentations. In the present study, a considerable variability was observed in the recording method of the clinical presentation of postaxial polydactyly of the hand in studies published previously. Thus, it is quite challenging to compare the clinical features among individuals of different racial and ethnic backgrounds. Therefore, unified recording methods must be developed to record clinical features so that they may be compared precisely. We would like to propose a recording method to record such clinical features, and an example of this type of clinical record is shown in Table 11. The classification systems that were used in the sample clinical record are shown in Table 12. This classification system combined the Temtamy-McKusick

Table 11 Clinical record of postaxial polydactyly of the hand.

Facility: maternity hospital general hospital others

Department: plastic surgery orthopedics pediatrics obstetrics others

Nationality: *Ethnic group:*

Sex: male female *Affected side:* unilateral (right left) bilateral

Family history (degree of consanguinity)

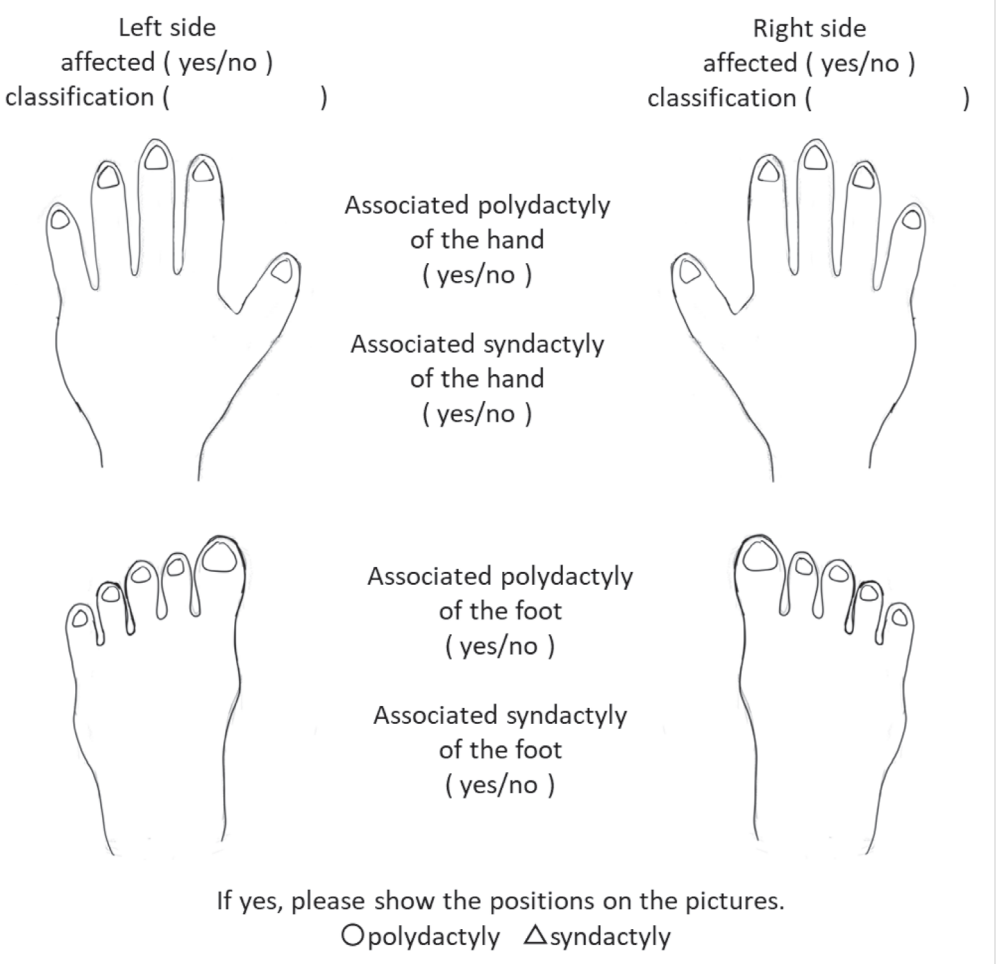
Postaxial polydactyly of the hand:

Other hand or foot abnormalities:

Treatment: observation ligation surgery

Associated syndromes: no yes:

Associated systemic abnormalities: no yes:



classification system and the Pritsch classification for type A polydactylies[19]. There are detailed classification systems for postaxial polydactylies of the hand, such as Stelling classification[2], Rayan-Frey classification,[1] and Pritsch classification for type A[19]. We used the Temtamy-McKusick classification system because of its simplicity and the difficulty in changing from this system to other classification systems. Though, this classification system is too simple, this is not adequate for a detailed classification. This was because we made the classification systems presented in Table 12.

Table 12 Classification system.

TypeA-1: metacarpal type; a fully developed sixth ray that articulates separately with the carpals
TypeA-2: metacarpophalangeal type; the extra digit occurs on the lateral side of the fifth digit with an intercalated distal metacarpal remnant
TypeA-3: phalangeal type; the supernumerary digit arises from a hypoplastic sixth metacarpal or is fused to the fifth metacarpal
TypeA-4: intercalated type; the extra digit originates from the fifth metacarpophalangeal joint
TypeA-5: fully developed type; the ulnar digit originated from a bifid fifth proximal phalanx
TypeB-1: tissue nubbin type
TypeB-2: connected with narrow skin bridge type

This classification system is a combination of Temtamy–McKusick classification and Pritsch classification.

Conclusion

In this study, the Japanese cohort with postaxial polydactyly of the hand may have clinical presentations that are unique and different from those of the Caucasian, African, and Middle Eastern cohorts. In the present study, it was challenging to compare accurately the clinical features of individuals of four racial and ethnic backgrounds because of inconsistent clinical records. A defined recording method to record the clinical presentations of such conditions must be developed for further investigations. In the future, we aim to further develop a recording method to record clinical features, and we suggest more detailed, large-scale international studies to be conducted.

Conflicts of interest

None declared.

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