

## The oldest reported mirror hand? The curse of the coal-house frog!

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### ABSTRACT

Despite being one of the rarest congenital upper limb abnormalities, a wide spectrum of the typical mirror hand has been described in the literature. We report a very interesting case of a new variant of mirror hand presenting in a 78-year-old man. The unique features of the case include the age at diagnosis, the anatomical features present and the acquired function despite no form of reconstructive surgery.

### KEYWORDS

Laurin-Sandrow Syndrome – Congenital Abnormalities – Polydactyly

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Mirror hand is one of the rarest congenital abnormalities affecting the upper limb, with around 70 cases reported in the literature. It is characterised by multiple digits, typically seven in total, with symmetry about the midline, absent thumb, excess carpal bones, absence of radius and duplication of the ulna (ulna dimelia).<sup>1</sup> We describe an unusual case of delayed diagnosis of mirror hand that presented to a hand unit when the patient was aged 78 years.

### Case history

A 78-year-old man was referred with a suspicious skin lesion on his left shoulder and at the time an abnormal left upper limb was noted. He was on dialysis for chronic renal failure, secondary to polycystic kidney disease. He was born via normal delivery to non-consanguineous parents with no family history of congenital defects. He had two fingers removed from his left hand at 6 months of age but no further investigation or reconstructive surgery had taken place. He was told by his mother that he was born with a hand like a frog as a consequence of a frog having jumped on to her abdomen during pregnancy while collecting coal from a coal-house!

On examination, the patient had absence of the thumb with five fingers on his left hand. The most radial digit tended to partial opposition to the other four fingers, similar to the appearance after a pollicisation procedure (Fig 1). He had a range of movement of 15–70° at the wrist and 10–60° at the elbow. The forearm was held in pronation and the left arm was slightly shorter than the right (Fig 2). An arteriovenous fistula was present to facilitate haemodialysis.

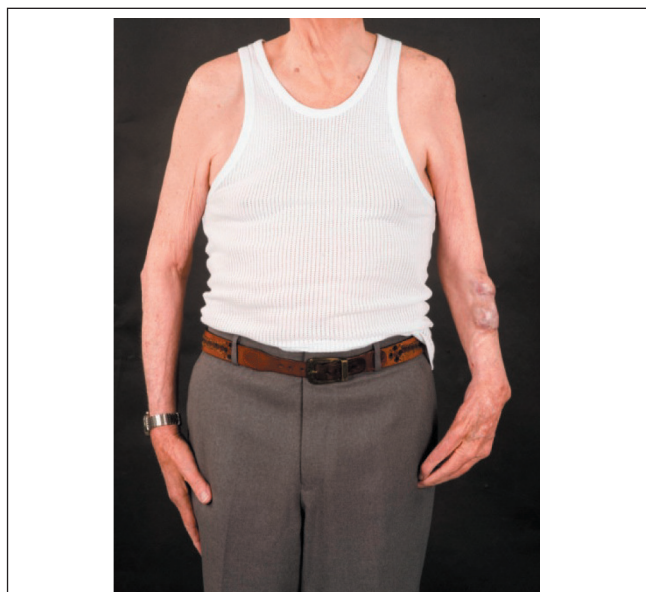
The patient had good function with both hands. He worked previously as a draftsman for a toy company and in

his spare time he was keen on DIY. He had no need to hold a pen in his left hand as he was right hand dominant but holding a knife and fork or other household objects caused no problem. He drove a non-adapted manual car.

Radiological evaluation revealed five triphalangeal digits with normal metacarpals and an absent thumb. The radius was absent and two ulna bones were present.



**Figure 1** Left forearm and hand: The posture of the most radial digit tends towards opposition.



**Figure 2** Photograph demonstrating the shorter left forearm and flexion deformity of the wrist. The arteriovenous fistula is evident.



**Figure 4** Vestigial humerus extending proximally from the lateral epicondyle to mid-humerus level



**Figure 3** Oblique and lateral plain x-rays of left wrist and forearm, showing duplicate ulna and additional carpal bones. Each ulna has an olecranon.

Interestingly, both ulnas had a broadened distal end, which mimicked that of distal radii (Fig 3). The humerus appeared abnormal with a thin vestigial bone extending proximally from the lateral epicondyle to the mid-humerus level (Fig 4). The scapula and humeral head were hypoplastic in comparison to the right. More detailed radiology could not be arranged as the patient passed away soon after presenting to our department.

**Table 1** The classification of the mirror hand-multiple hand spectrum described by Al-Qattan<sup>2</sup>

Type	Features	Syndrome
1A	Ulna dimelia (classic mirror hand)	
1B	Ulna dimelia – preaxial ulna hypoplastic	
2	2 ulnas (one vestigial) + radius	
3A	Multiple fingers + 1 ulna + 1 normal radius	
3B	Multiple fingers + 1 ulna + 1 hypoplastic radius	
4A	Bilateral multiple fingers with complex syndactyly + feet/facial defects + ulna dimelia	Sandrow
4B	Bilateral multiple fingers with complex syndactyly + feet/facial defects + ulna + radius	Martin
5	Multiple hand (complete duplication) + normal ulna/radius	

**Discussion**

A wide spectrum of variants ranging from ulna dimelia to the multiple hand have been described in the literature. Al-Qattan produced a classification of the mirror hand-multiple hand spectrum (Table 1).<sup>2</sup>

The exact genetic basis is unknown. However, the most commonly proposed event is impairment of the mechanism responsible for the organisation and differentiation of the limb bud,<sup>5</sup> not that of primary duplication, which was initially thought to be the case. Several authors have demonstrated cases where the distal end of the preaxial ulna has broadened to take on a shape similar to that of the radial head, to aid mechanical function of the hand, which tends

to favour failure of differentiation as the primary event.<sup>2-4</sup> In our case, both ulnas had a broadened distal end, similar to that of a distal radius. To our knowledge, this is the first description of such a case in the literature, possibly due to the age of the patient at diagnosis, resulting in a protracted period for bony remodelling. This may possibly be an adaptation to the abnormal mechanical load at the wrist between the two ulnas and the abnormal carpus.

It is unknown whether the patient's renal disease was associated with the congenital hand. However, there is one reported case in the literature of a child with mirror hand who also had a multicystic kidney, raising the possibility of a link.<sup>5</sup>

Treatment options vary between the spectrum of defects. The main principle of surgical correction is amputation of the accessory digits (usually radially), either at the level of the carpometacarpal joints or at the metacarpal base, and pollicisation of the most radial retained digit. Problems with flexion deformities of the wrist can be treated with splinting, arthrodesis of the joint or tendon transfer to aid wrist extension. Our case highlights that it is possible for the upper limb to adapt to provide a well functioning hand, even in the absence of reconstructive surgery.

## Conclusions

We describe a case of mirror hand in a 78-year-old man, who despite no reconstructive surgery had a well functioning hand. Not only is this case unusual due to the age of the patient at diagnosis but also because of the features present. We believe our patient may be a new variant on the typical 1A mirror hand, with duplicate ulnas, multiple metacarpals and fingers, associated with an abnormal distal humerus, which we believe has not been described previously in the literature.

## References

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