CONGENITAL DEFORMITY OF THE CARPUS ASSOCIATED WITH
MALDEVELOPMENT OF CERTAIN THENAR MUSCLES

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In 1943 Hodgson described a case of "congenital retardation in development of the carpal navicular, first metacarpal and styloidal process of the radius" in association with wasting of the thenar muscles and recurrent sprains of the wrists. He was unable to find any previous description of a similar case and no other seems to have been recorded since 1944. The present paper describes two further patients with this condition, each of whom had additional congenital abnormalities.

CASE REPORTS

Case 1—A woman of thirty-one was admitted primarily for mitral valvotomy, and attention was drawn to the deformity of her hands by the finding that arterial pulsation at the wrists was greatly diminished. Both hands had a characteristic rather simian appearance (Fig. 1). There was marked radial deviation at the wrists, the movements of which were, however, painless and of normal range. The thumb was abnormally long and slender and resembled an extra finger, this appearance being accentuated by the almost total absence of the thenar muscular eminences. The power of abduction of the thumb (at right angles to the palm), of opposition of the thumb and of flexion of the first metacarpal-phalangeal joint was greatly reduced, and the muscles concerned—abductor pollicis brevis, opponens pollicis and flexor pollicis brevis—gave a negligible response to faradic stimulation. On the other hand the adductor pollicis was present and of normal strength, as were the first and other interosseous muscles. The hypothenar muscles were present, though slightly underdeveloped. There was no apparent wasting of the muscles of the forearm or upper arm. The supinator, biceps and triceps reflexes were present and equal on the two sides, and there was no loss of sensation. Systemic neurological examination revealed no abnormal signs. Arterial pulsation could not be felt in either radial artery, and only a very feeble pulsation was detectable in the ulnar vessels, whereas brachial and axillary pulsation was easily felt. Arterial pulsation at the wrist was not increased by elevating the arm. Nothing abnormal was palpable at the root of the neck.

The patient said that her hands had been "queer" all her life, but that they had caused her little disability apart from slight weakness of the grip. She had not suffered any injury to the wrists, and had not been subject to sprains. She had had seven children, none of whom is, so far as she can tell, similarly affected (they were not examined by us), and she knows of no comparable case amongst her relations.

Radiological examination. Left hand—The scaphoid bone was strikingly smaller than normal and had a sharply defined elliptical shape quite unlike the normal scaphoid (Fig. 2). The density of the bone was slightly increased. The proximal end articulated with the lateral aspect of the articular surface of the radius, but a wide gap separated the distal pole from the capitate and trapezium. The styloidal process of the radius was poorly developed, the articular surface
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FIG. 1
Case 1—Right hand shown with a normal hand for comparison. The hand and arm of the patient were in the most comfortable position; the normal hand was not.

FIG. 2
Case 1—Radiographs of the hands.
of the lower end of the radius being set at right angles to the long axis of the shaft, so that the ulnar and radial styloids lay at the same level. The base of the first metacarpal bone was rounded and showed neither an abductor tubercle nor the customary saddle-shaped articular surface. In consequence the first carpo-metacarpal joint was very shallow. The shaft of the first metacarpal bone was unusually elongated and slender. A single sesamoid bone was present at the first metacarpo-phalangeal joint. Right hand—In this hand no separate scaphoid could be identified but the articular surface of the radius was irregular and the appearances suggested that a deformed and rudimentary scaphoid was joined with the lower end of the radius (Fig. 2). In addition there was fusion of the capitate and hamate bones. The first carpo-metacarpal joint was shallow, and the first metacarpal bone was unduly long and thin, as in the left hand. Feet—Both feet showed conjunction of the cuboid and lateral cuneiform bones. Thoracic inlet—Large bilateral cervical ribs were present.

Case 2—A man of sixty-three presented with a complaint of pain in the legs and difficulty in walking. In the course of neurological examination his hands were noted to have a curious simian appearance and there was almost complete absence of the thenar eminences. There was, however, no wasting of either the first interosseous or adductor pollicis muscles and the other intrinsic muscles of the hands also appeared normal. In spite of the apparent absence of the opponens pollicis muscle his grip was fairly strong. Neurological examination showed no other abnormality: the deep reflexes of the arms and legs were normal and there was no sensory loss. He said that the deformity of his hands had been noticed in early childhood and that it had caused him no disability, although he had worked as a labourer for many years. He had never had pain in the arms or shoulder girdle, nor had he ever had a fracture or sprain of either wrist so far as he could remember. He knew of no similar condition among his relations.
Radiological examination—In the left carpus the scaphoid was small and elliptical and lay transversely, its non-radial aspect being widely separated from the capitate bone and trapezium (Fig. 3). The general appearance of the scaphoid in this instance closely resembled that of the left side in Case 1. The right carpus was radiologically normal. Cervical spine—There was a spina bifida of the first thoracic vertebra (Fig. 4) and some degeneration of the cervical intervertebral discs, but no cervical ribs were present. Intravenous pyelogram—This showed crossed renal ectopia, both kidneys lying on the right side of the abdomen.

DISCUSSION

The two patients described were very like the patient in Hodgson's (1943) case in that they had a selective wasting of the thenar muscles in association with a carpal deformity which involved principally the scaphoid bone, and it seems probable that these lesions comprise a distinct entity. Unlike Hodgson's patient, neither of our patients had suffered much disability from the muscular lesions in spite of the almost total absence of opponens pollicis, a muscle that is of major importance in the power of the grip. It is difficult to be sure whether the affected muscles were wasted in the true sense or were in fact congenitally absent or maldeveloped. Both of our patients had lesions of the cervico-thoracic vertebrae which might conceivably have accounted for the muscular deficiency. Thus the patient in Case 1 had bilateral cervical ribs, and the combination of obliteration of the radial pulses with wasting of the thenar muscles might have suggested that the cervical ribs were responsible for both lesions. Nevertheless the complete absence of pain or sensory loss, and the presence of the muscular deficiency from early childhood, make it highly unlikely that the cervical ribs were in fact culpable. Similar arguments may be applied to the spina bifida in Case 2. All three
of the affected muscles of the thumb are supplied principally by the median nerve, and although it is possible that a lesion of a branch of the median nerve may have been the cause of the muscular lesions this is also unlikely in view of the total lack of sensory impairment.

Whatever the etiology of the muscular lesions there seems to be no doubt that the carpal deformity is of congenital origin. As with most congenital anomalies it is not possible to give a certain explanation of the origin of the lesions, but it is perhaps of interest that Wood Jones (1941) believed that "the human scaphoid is a compound bone, being composed of the primitive scaphoid in its proximal part and the pre-axially displaced os centrale in its distal part." In Figure 5 we reproduce Wood Jones's diagram, comparison of which with the radiographs of the left scaphoid bones in Cases 1 and 2 suggests that the deformity may possibly represent persistence only of the proximal part of the scaphoid, unfused with the distal part. Even if this were the correct explanation, however, it would not account for the other changes in the carpus, although the abnormalities of the first metacarpal might be related to the absence of its normal muscular attachments. It is difficult to envisage any direct relationship between the carpal deformity and the muscular deficiency, because none of the muscles involved has any material attachment to the scaphoid bone.

It seems probable that the condition described is less rare than the paucity of recorded cases would suggest, because it may cause relatively trivial symptoms and thus escape detection. In obscure cases of "wasting" of the intrinsic thumb muscles it may be worth while to radiograph the carpus.

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REFERENCES
